



Caring for Children with Chronic Health Conditions

A Guide for Intact Family Agencies

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OFFICE OF THE
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PREFACE

This guide was developed to assist DCFS-funded Intact Family Agencies in their work with parents and relatives caring for a child with a chronic or complex health condition. The family may have been the subject of a medical neglect investigation and referred for intact services. In these cases, it is critical that the Intact Family Worker gains an understanding of psychosocial, cultural, community and environmental factors that potentially and actually influence the family's care of a child with a health condition. Intact Workers may need to help the family lower the potential risk of ineffective health management by assuring that the family has an adequate comprehension of the illness to make informed choices, help the family overcome a feeling of powerlessness because of complex treatment regimens and decisional conflicts that can occur between a system of healthcare providers and the family. Risk prone health behavior may be related to an array of factors such as: inadequate comprehension of the illness, economic disadvantages, insufficient resources, lack of means of transportation, stressors, role strains, insufficient or perceived barriers to social support, exhaustion and a myriad of ineffective tension release strategies (Herdman & Kamitsuru [Eds.], 2014/2017).¹ Knowledge of and seeking the assistance of advocacy agencies may help the family obtain needed resources.

In cases involving infants, Intact Workers need to be facile in their communication and linkage to local and state public health and other community resources. Obtaining consents to communicate with APORS and WIC services at case opening may prevent missed appointments for vulnerable infants. Additionally, Intact Workers need to obtain consents from the parents for the Intact Workers to communicate with the child's medical home regarding the child's health and medical care management. In cases where a child is prescribed medication, the Intact Worker should monitor and document the use of the medication on a weekly basis. The introduction section of this manual is designed to assist the worker in discussing topics relevant to the individual family. For example, if the family appears to be hesitant in asking for support from their extended family or informal support systems, the section on Friends and Family may assist the family overcome their reluctance. The relevant sections to the manual can be printed out and shared with the family for discussion purposes. The caseworker is free to use those sections of the guide that are related to the family's individual circumstances. This manual also includes specific information about six specific chronic health conditions, however the general principles included herein apply to all chronic health issues.

¹ Herdman & Kamitsuru (Eds.) (2017). Nursing Diagnoses (ND). In S. I. S. Will, M. W. Arnold & D. S. Zaiger (Eds.), *Individualized healthcare plans for the school nurse* (pp. 84-86). Forest Lake, MN: Sunrise River Press. (Original work published 2014)

CHAPTER ONE: INTRODUCTION

Caring for a child with a chronic illness adds to the challenges of being a parent. There can be added appointments to juggle, or special diets and medication schedules to follow. It can add unpredictability to life, and even cause stress on the entire family. Navigating care for a child with a chronic illness is difficult, but it is doable. The more you know about a chronic condition, the better you'll be able to make a plan for care, anticipate challenges down the road, and know where to look for help.

This introductory chapter to the guide will help you understand how to care for a child with a chronic condition. First, you'll be introduced to the types of knowledge needed to care for a child with a chronic condition – general knowledge, specific knowledge, child knowledge, and family knowledge. Second, this chapter will help you to channel that knowledge into a plan tailored to your child that maps out available resources for providing care.

At the end of this chapter, you will find a case vignette and questions to help facilitate discussion for case managers and supervisors about how to better manage cases involving children with chronic illness.

TYPES OF KNOWLEDGE

GENERAL KNOWLEDGE

General knowledge means understanding what a chronic health condition is: a health problem that lasts for a long time, usually more than three months. When a child has a chronic health condition, they need special care and attention from the parents, family and friends, health providers, and school, social and community services.

“Chronic health condition” is an umbrella term that describes several long-term health problems that impact different parts of the body. This guide covers asthma, diabetes, sickle cell disease, cystic fibrosis, cerebral palsy, and seizure disorders, but there are many others, such as HIV, or even cancer.

Over time, a chronic health condition can impact many parts of a child's life in ways that extend into adulthood, including how a child feels both physically and emotionally, develops, moves, and talks. In turn, a child's chronic health condition can affect the entire family.

It's important to understand that people with a chronic condition can be well at any given time, just as the condition can cause them to be sick. Whether a child with a chronic condition is healthy or ill, they always have that chronic condition. The goal in managing and treating a chronic condition is to control the overall effect of the condition so that a person can live the healthiest, safest, and happiest life possible.

SPECIFIC KNOWLEDGE

One of the most important parts of making a plan of care for a child with a chronic condition is to gain specific knowledge about that condition. There are a lot of different chronic health conditions, and each has its own causes, symptoms, management and treatments. For example, caring for someone with asthma is very different than caring for someone with Cerebral Palsy or Diabetes.

In addition, each chronic condition has different levels to the disease. For example, some children with asthma, a very common condition, have mild symptoms, and may not even need to take medications

every day. Other children with asthma have severe symptoms that can sometimes be life-threatening, and need to take multiple medications every day.

The more specific knowledge you have about your child's chronic condition, the better you'll understand how the condition impacts your child and plan for the care your child needs.

CHILD KNOWLEDGE

Now that you've learned about general and specific knowledge, it's time to talk about child knowledge—how a chronic condition impacts your child specifically. The effects your child may experience can be sorted into 3 categories: Physical, Cognitive and emotional.

PHYSICAL

A chronic condition will physically impact your child uniquely. For example, children with epilepsy, a disorder that causes seizures, may have seizures in different ways. Getting to know how your child's condition "shows" itself—what your child looks like, how they behave or what they say when they're having symptoms or getting sick—is a very important part of making a plan to keep your child safe and knowing when to take action.

Additionally, depending on the chronic condition your child has, your child's physical growth may be affected. And sometimes, special diet or treatments for a condition may impact how your child develops. Complications from a condition, including problems with fighting off infections, or healing from injuries, can also affect how your child develops. Understanding how your child's chronic condition impacts their physical development will help determine what kinds of special services and supports they may need.

COGNITIVE

Some chronic health conditions don't affect how a child thinks. However, some chronic conditions do, and can change the way your child thinks and solves problems. Knowing whether and how a condition impacts your child's cognitive thinking is important in making a plan to make sure your child is accommodated at school and has access to programs and supports that help them succeed to their highest potential.

EMOTIONAL

No matter the type, having a chronic condition almost always emotionally impacts a child. Unlike children who get sick with a cold or the flu, kids with chronic health conditions must cope with the fact that their illness is here to stay, and may change their lives in big ways.

The emotional response children have in response to a chronic illness is sometimes referred to as "trauma", and is different for each child depending on their personality, resiliency, environment, and the specific illness they have. One major factor in how children process their condition is their age. For example, young children often cannot understand why they have an illness. They may think it's a punishment, or they may get angry at their parents, caregivers, or doctors for being unable to cure them. As children grow older, they may better understand the causes of their condition, but may become frustrated and angry, especially if they feel that their chronic illness is causing them to lose their independence or to be left out.

At any age, a chronic health condition may cause your child to think negatively of themselves. This can be worsened if people stare at, make fun of, or exclude your child from activities at home or school because of their condition.

It's important to pay close attention to how your child is coping with their chronic illness. The more you learn about how your child is impacted emotionally, the better you'll be able to support them. It will also inform discussions and decisions on whether your child needs emotional counseling or other supports.

FAMILY KNOWLEDGE

The last type of knowledge related to your child's chronic condition is family knowledge: how a chronic condition impacts you, other members of your family, and your family as a whole.

SIBLINGS

One of the most important parts of family knowledge is understanding how siblings of a child with a chronic condition are affected. Although having a sibling with a chronic condition is different than having a chronic condition itself, it can still cause stress. Children can react in a lot of ways, depending on age, type of condition, and the relationship the sibling has with the child with the condition. They may feel:

- Worried about their sibling
- Jealous of the attention their sibling gets as a result of the condition
- Upset that they can't go certain places or do certain things because of their sibling's illness
- Embarrassed that their sibling has a chronic condition
- Responsible for their sibling having a chronic condition
- Guilty that their sibling has a condition

Paying attention to how each of your children reacts will help you plan for any supports they need, whether it's just setting aside special time for them, helping school staff support your child, or accessing counseling to help your child cope with their sibling's chronic illness.

PARENTS AND CAREGIVERS

Parents of children with chronic conditions have a lot to keep track of. From understanding the condition and making sure your child keeps up with treatment and appointments to knowing how it affects the child and their siblings, the responsibilities can add up to a lot of stress!

That's why it's important to know how you and any other caregivers are impacted by caring for the child. In addition to stress, parents and caregivers may feel sadness or grief for the child, or even frustration and resentment. Certain parts of caring for a child with a chronic condition may be particularly stressful or exhausting, while others come easily.

Knowing how a child's chronic condition affects you will help you to know when to ask for help, seek support, or find someone to watch your children when you need a break.

FURTHER READING ON CARING FOR CHILDREN WITH CHRONIC CONDITIONS

Children with Chronic Conditions

University of Michigan

<http://www.med.umich.edu/yourchild/topics/chronic.htm>

Caring for a Seriously Ill Child

Kids Health

<http://kidshealth.org/en/parents/seriously-ill.html#>

MAKE A PLAN WITH YOUR PARTNERS IN CARE

Now that you've learned about the four types of knowledge related to chronic health conditions, you can start to learn how to make a plan. A plan for caring for your child should address the needs you, your child and family may have: for taking medications regularly; for acting if your child becomes sick; for getting to doctor appointments; for helping teachers and others at school understand and care for your child; for making sure all other family members are taken care of, including yourself.

In making your plan, it's important to understand that you should not be alone in executing it. You have many partners in care that can help you make and execute your plan. You need to know who your partners are, what they do, and when to use them. For example:

- Your child's primary care provider, local hospital staff, nurses or social workers will help determine the medical treatment necessary for your child, and teach you about how to properly administer it and monitor for signs that your child may need urgent medical attention or changes in long-term treatment.
- Staff at your child's school will administer any medications your child may need and make sure they stay safe during the school day.
- Members of your family or friends or neighbors might care for your child, help you and your child get to appointments, or watch your other children while you're away.
- Resources in your community can provide you, your child, and family extra support.

It's also important to know that sometimes, especially when caring for a child with complex health needs, things just go wrong. You might have trouble refilling a prescription for your child's medication. A ride to your child's doctor appointment might cancel or show up late, and your arrangement for taking care of your other children while you're away might fall through. You may get frustrated, sometimes to the point where you feel like you can't do it anymore and want to quit.

Your partners in care can help in these situations too. Any time your plan goes off the tracks, the most important thing is to communicate with your partners about the difficulties you're having. A big part of their jobs is to help you navigate what to do and come up with solutions when things go wrong.

It is hard to ask for help. Just remember, you are not asking people to help you; you're asking people to help your child.

In the next four sections, you'll be introduced to partners in care from four places you'll likely work with in making your plan for care: school staff, health professionals, friends and family, people who provide community resources.

SCHOOL STAFF

In making a plan for care for your child, you'll work with multiple partners at your child's school to make sure that the safety and health needs of your child are met at school the same as they are at home. These partners include your child's teacher, school nurse, social worker and principal.

Once you've informed your child's school that they have a chronic condition, you'll meet with your partners in care at school to craft a plan for your child when they're at school.

The plan will draw a lot from your child knowledge and specific knowledge, and should include:

- Your child's medical history
- Your child's special needs
- Medicines or procedures your child needs during the school day
- Special dietary needs your child has
- Your child's transportation needs
- Possible problems your child may have as a result of their chronic condition, and precautions the school should take to avoid them
- Your child's pediatrician's name and contact information
- Emergency contacts, including you, your child's pediatrician, and someone you trust that the school can call if you're not available.
- Plans for what to do in an emergency.
- Special child knowledge you have that can help school staff calm or soothe your child in stressful situations, identify signs that your child is getting sick, and avoid any triggers that could set your child off.

Your partners in care at school will also work with you and your child to make sure that your child can learn to the best of their ability, while considering your child's medical needs and how they can impact learning. There are laws to make sure that this happens, and the process can include a specialized plan for your child, known as Individual Education Program Plans (IEP) or Section 504 plans.

Making a care plan for your child at school can be overwhelming. It's good to remember that most staff members have experience working with children with chronic conditions, and will help guide you through the planning process. In addition, here are some pointers to stick to and think about along the way:

- Talk to your partners at your child's school. Don't be afraid to talk to school staff about your child's condition. Although it's natural to be worried about sharing this information, the more you share, the better staff will be able to provide care, and the more your child will be able to learn and thrive at school. Without information, staff members may make false assumptions about your child.
- Seek out a school social worker. Ask if your child's school has a social worker. If they do, ask to meet with them. More than any other partner in care at school, social workers may be especially helpful arranging for transportation or special educational accommodations.
- Plan Ahead. Plan to meet or talk regularly with your child's teacher and always ask if your child's condition is affecting behavior or schoolwork. If your child misses class frequently due to a chronic condition, make a plan with their teacher to make it up.
- Make sure medications are stocked. Communicate with the school nurse to make sure that medications and other medical supplies your child needs never run out.
- Plan with your child. If possible, talk through the school-day routine with your child, including when and where they should take medications, what to be careful about, and who they should go to for help, or what to do if they feel sick.
- Inform the teachers of your other children that their sibling has a chronic condition. This is especially important if your child's condition has impacted your other children emotionally or behaviorally. Talking to their teachers will help them to understand why your children may be behaving differently from what they expect, and know how to better support them.

MEDICAL PROFESSIONALS

Every medical professional partner in care that you work with will help you gain general and specific knowledge about your child's chronic health condition. In turn, you'll give each health professional child knowledge to help them understand how your child is doing with the condition overall or reacting to a treatment. By doing this, you and your child's health professional will each contribute to a plan for medical care that best fits your child's needs.

There are many types of medical professionals you might work with in caring for your child. For example:

- You'll work with a primary care provider – either a pediatric doctor, nurse practitioner or physician's assistant who specializes in caring for children.
- Depending on your child's medical needs, you might work with specialist providers who treat certain parts of the body or conditions.
- You might work with nurses to learn how to manage your child's condition day-to-day, or physical, occupational and speech therapists, who help with movement, daily life activities, and speaking.
- You and your child might work with a social worker in addressing emotional needs, or a nutritionist, for advice on following a special diet.

Depending on your child's needs and where you live, your child might get healthcare at a medical home, a model for primary care that coordinates health management across specialists, hospitals and home.

NAVIGATING A DOCTOR'S VISIT

Doctor's visits can be overwhelming. Often, just getting there can be a burden, but usually there's a lot more involved. You have to know how your child is doing, the medications they're taking, and keep track of all of the concerns you or your child have had since the last visit.

In preparing for a doctor visit, it's good to make a checklist of things or information to make sure you have. This includes:

- A list of the medications and supplements that your child is taking
- A list of your child's allergies
- A copy of your child's immunization records
- Knowledge of how your child is doing. This includes any unusual changes in how your child feels or in their behavior. Try to take note of when this happen, and let the doctor know if you think there's anything that causes this change.
- Summaries of visits from any other specialist or health professional your child sees for their condition.

You might find it helpful to keep all of these documents together in a folder at home, so that, when needed, you can just grab the folder and not have to search.

Once you're at the appointment with your child, it's also important to make sure your child's provider has what they need to take good care of your child.

This includes your child's medical records from:

- Previous primary care providers your child may have
- Visits to specialists
- Diagnostic testing
- Any past hospitalizations

Knowing which questions to ask during an appointment can be overwhelming in and of itself. Appointments sometimes move fast and can feel rushed. It's important to remember that you have the right to have all of your questions during each appointment with a medical professional. No question you have is unimportant, and it's a good strategy to keep asking question until everything about your child's health and the plan going forward is clear to you. Here are some questions to address at each visit:

- How is my child's health overall?
- What do different tests show about my child's condition?
- Why is my child taking each currently prescribed medication?
- What goals would you like to see my child reach by the next visit?
- Where can I go to learn more about my child's condition?
- What problems might happen to my child that would make me call 911 for emergency care?
- How often should we be making a visit to see you?
- Who should I contact if something goes wrong or I have questions and what is their contact information?
- Is there a social worker here that might be able to help if I am having trouble getting to appointments?

As the appointment is nearing its end, the most important thing to understand is the plan for next visit. This includes:

- Any medications your child is starting or stopping, and the medications they're continuing
- Any tests that your child will receive before the next visit, and when he or she will receive them, and how to make those appointments.
- Any specialists your child will see before the next visit.

NAVIGATING DIFFERENCES OF OPINION BETWEEN MEDICAL PROVIDERS

If your child sees multiple providers, it's possible that at some point, you'll receive conflicting advice. For example, your child's pediatrician could recommend one medication, while their neurologist recommends another. This is normal – differences in opinion happen all of the time. But it's important that the difference be resolved as soon as possible so that you don't have any confusion about what the plan of care for your child should be.

If you notice a difference in opinion, make both providers aware of the conflict and ask them to talk to resolve it. This is often the easiest solution.

If they continue to disagree, or won't communicate with each other in a timely manner, reach out to your caseworker, and ask for a conference between the doctors and the DCFS Medical Director to resolve the issue.

NAVIGATING DIFFERENCES OF OPINION BETWEEN YOU AND MEDICAL PROVIDERS

Just like disagreements between providers, there will be times when you won't agree with the care plan that a medical provider would like you and your child to follow. This is also normal, but it should be addressed with the provider you have a disagreement with as soon as possible. If you don't, the provider may challenge you, especially if they feel that the child could be hurt from not getting the care or treatment they feel the child needs.

If you encounter a disagreement with your child's medical provider, it's important to keep a few things in mind:

- The provider knows you're acting in the best interest of the child.
- The provider understands that you know your child best, and that you know how decisions for your child will affect you and your family.
- The provider has a lot of medical knowledge about the condition your child has that can sometimes inform recommendations for treatment that are different than your own.

Keeping these things in mind, the best way to resolve a disagreement with a medical provider is with direct communication. Share your feelings and concerns with the provider. Ask questions. Often, disagreements come from misunderstanding on both sides.

If this doesn't work, talk to your caseworker about it, letting them know your concerns and why you disagree with your provider. Your caseworker will likely refer you and your doctor to the DCFS Medical Director to settle the disagreement.

TROUBLESHOOTING MEDICAL CARE

There are a lot of moving pieces when it comes to caring for a child with a chronic condition. Almost nothing always goes right all the time, especially related to medical care. Sometimes, even the best laid plans fall apart.

When arranging, and getting medical care for your child, there are all kinds of things that can go wrong, such as:

- You lose transportation to get your child to an appointment
- A medication your child's medical provider prescribed is not covered by your insurance
- You lose your child's medical card
- Medical equipment that would improve your child's quality of life is too expensive
- You need care for your other children

Things go wrong, and your partners in care are there to help. And just as with resolving disputes, communicating to your partners and asking for help is the first and most important step in getting things to go right. Here are some examples:

- If you need a way to get you and your child to an appointment, call your child’s doctor’s office to notify them of your situation. A lot of doctor’s offices have someone on staff who can help you to brainstorm a solution.
- If a medication prescribed to your child isn’t covered by insurance, call your child’s doctor’s office. Often, there are alternative medications that can be prescribed, or they can lobby on your behalf with the insurance company.
- If your child needs a piece of medical equipment that’s not covered by insurance and is too expensive to pay out of pocket, talk to your caseworker. Depending on the situation, there are state funds that can be accessed to pay for such expenses.

FURTHER READING FOR UNDERSTANDING THE MEDICAL SYSTEM

Defining the Medical Home

Patient-Centered Primary Care Collaborative
<https://www.pccpc.org/about/medical-home>

Defining the PCMH

Patient Centered Medical Home Resource Center
<https://pcmh.ahrq.gov/page/defining-pcmh>

FRIENDS AND FAMILY

Your family and friends can be great partners in planning for and delivering care to your child. There are a lot of ways that they can help, both in situations that are regular and planned, and emergencies, when an arrangement falls through and you need someone to step in for extra assistance.

Unlike working with your school and medical professional partners in care, the role of your friends and family in making and executing your care plan are less set in stone. It’ll depend first on what you, your child’s, and the rest of your family’s needs are—informed by your specific, child and family knowledge. Second, it’ll depend on the capacity of your friends and family to help.

There’s a lot to consider when thinking about incorporating your friends and family into your care plan for your child. Here are four steps to help break down the planning into manageable pieces.

DETERMINE YOUR FAMILY’S NEEDS

What are your, your child’s and family’s needs in caring for a child with a chronic illness that you need help with? This can be a lot of different things, such as:

- Help getting you and your child to appointments
- Giving reminder calls for upcoming appointments
- Babysitting your children when you have other commitments or when you just need a break.

It’s also a good idea to think about what your needs might be in an emergency:

- Who can take care of your other children if your child with a chronic condition suddenly becomes sick and you have to take them to the doctor’s appointment or a hospital?
- Who else can you call if a child care arrangement falls through?

Once you have some ideas, try to make each idea as specific possible and make a list.

ASK YOUR FRIENDS AND FAMILY FOR HELP

Which members of your family and friends can help, and how? Different friends and families can help with different things. One family member might be able to drive you and your child to doctor's appointments, but they might not be the best at babysitting your children while you're away.

It's important to think about a person's dependability. Here are some questions to consider:

- Can you expect them to arrive on time?
- If you're asking them to care for any of your children, will they take good care of your children?
- Will they make sure that your children get to do their daily activities?
- Do they understand that children behave differently at different ages?
- Will they use non-violent and non-physical discipline strategies if your children misbehave?

If you're considering asking someone to help care for your child with a chronic condition, here are some additional questions to think about:

- Does that person understand the duties in providing good care?
- Can you count on your friend or family member to give your child their medications on time? If medications need to be given to your child a special way, can you count on that person to do so correctly?
- Can you count on the person to follow a plan in case of a medical emergency?

Sometimes, it can help to have a meeting with your friends and family to talk about you, your child's and family's needs and to ask for their help. Talking to family members and friends one-on-one is okay too. Asking for help can be difficult. Again, remember that you're not asking for help for yourself; you're asking for help for your child and family, who your friends and family all love and care for. Start by explaining that, while you're happy to care and be responsible for your child and family, there can be too much involved for one person to do it alone, especially when caring for a child with a chronic illness. When asking each family member and friend for help, be very specific about exactly what you're asking that person to do. No matter what, make sure that they understand what you're asking them to do, and that they're ready to make a commitment.

MAKE A PLAN

When you, friends, and family have agreed on a plan, write it down and give a copy to everyone who is agreeing to help. This will help you, your friends and family members keep track of who's doing what.

If any friends or family members agree to help care for your child with a chronic condition while you're away, this plan should include your child's special needs and the routine for care you follow during the day, including:

- General information about your child's chronic condition.
- Any medications your child takes, and when and how they take them.
- Information on any special diet that your child may have to follow.
- Possible problems your child may have because of their chronic condition, and precautions to take to avoid them.
- Your child's pediatrician's name and contact information.
- Emergency contacts, including you, your child's pediatrician, and someone you trust that the school can call if you're not available.
- Plans for what to do in an emergency.
- Special child knowledge you have that can soothe your child in stressful situations, identify signs that your child is getting sick, and avoid any triggers that could set your child off.

CHECK IN

Once your friends and family are helping, regularly check in with them to make sure that things are going well and that they're still happy to help.

This is especially important if you have an older child that's helping out. Siblings of children with chronic conditions can often feel pressure to grow up too fast and may come to feel upset or resentful if they're asked to help too much. Regularly checking in with any children that are helping out is central to avoiding these kinds of conflicts.

FURTHER READING ON CHILD AND FAMILY COPING

Caring for Siblings of Children with Special Needs

Kids Health

<http://kidshealth.org/en/parents/siblings-special-needs.html?WT.ac=ctg#catfamily>

Families Taking Charge: Dealing with the Stress

Virginia Cooperative Extension

<http://www.pubs.ext.vt.edu/2811/2811-7016/2811-7016.html>

Supporting Children with Chronic Illnesses

Pediatric Cardiac Society of South Africa

<http://www.pcssa.org/supporting-children-with-chronic-illnesses/>

Coping with a Chronic Illness

Lutheran Health Network

<http://www.lutheranchildrenshosp.com/coping-with-a-chronic-illness>

Siblings of Kids with Special Needs

University of Michigan Medicine

<http://www.med.umich.edu/yourchild/topics/specialneed.htm>

Children with Chronic Illness: Dealing with Emotional Problems and Depression

Healthy Living

<https://www.healthychildren.org/English/health-issues/conditions/chronic/Pages/Children-with-Chronic-Illness-Dealing-with-Emotional-Problems-and-Depression.aspx>

COMMUNITY RESOURCES

The Arc of Illinois

20901 LaGrange Road Suite #209

Frankfort, IL 60423

(815) 464-1832

<http://www.thearcofil.org>

Early Intervention Programs for Infants and Toddlers with Disabilities: 0 - 3

Illinois Department of Human Services

(800) 447-6404

<http://www.dhs.state.il.us/page.aspx?item=303>

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Crisis Mental Health Services for Children and Youth

Screening, Assessment, and Support Services

Illinois Department of Human Services

<http://www.dhs.state.il.us/page.aspx?item=92597>

Division of Developmental Disabilities

Illinois Department of Human Services

(888) 337-5267

<http://www.dhs.state.il.us/page.aspx?item=322>

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CASE VIGNETTE

A nine-year-old central Illinois child who suffered from Rett Syndrome², a neurological disorder, died. The medically complex child was hypotonic, non-verbal, non-ambulatory, and received all nutrition through g-tube feedings five times a day. She was dependent on others for all of her daily care, including toileting. She was also diagnosed with a seizure disorder, common in patients with Rett Syndrome. Children with this syndrome often develop autistic-like behaviors, breathing irregularities, feeding and swallowing difficulties, growth retardation, and seizures. Death is often related to seizure, aspiration pneumonia, malnutrition and accidents. Approximately one quarter of deaths from the Syndrome are sudden. An autopsy conducted after the child's death determined that she died from cardiac arrhythmia due to Rett Syndrome.

Three months prior to the child's death, an intact case was opened after the child's mother was indicated for medical neglect. Pharmacy records revealed that the mother had failed to consistently refill her daughter's primary anti-seizure medication. The child had been prescribed the anti-seizure medication by a pediatric neurologist from a children's hospital in Chicago. The mother and child had travelled to Chicago because there was no pediatric neurologist in the area where they resided. A few months later the child was seen locally by another pediatric neurologist who had recently relocated to the area. This physician prescribed a secondary medication to help control the seizures. The child's pediatrician who had followed the child since infancy opined that the secondary medication was not essential and could be used on an "as needed" basis. Consequently, the mother rarely administered the secondary medication. Later during the child welfare investigation after the child's death, the local pediatric neurologist took issue with the child's pediatrician's position regarding his opinion that the secondary medication was an "as needed" medication.

An Intact service caseworker was assigned to assist the mother obtain supportive services and monitor medication compliance. The single mother who worked in manufacturing admitted to being overwhelmed caring for her daughter and two other children and expressed frustration over her lack of health insurance coverage for medical equipment such as a lift to help bath her daughter. The worker offered to contact the child's insurance provider to request home nursing assistance and medical equipment but the mother assured the worker that her effort would be in vain as she had requested services numerous times and had been denied.

The worker who had no prior experience with Rett Syndrome, familiarized herself regarding the disorder through a Google search. When interviewed, the mother minimized her daughter's seizure disorder and maintained that the child did not suffer from seizures but rather had tremors. The mother explained that while having a tremor, her daughter would remain alert, thereby distinguishing this behavior, in her opinion, from a seizure. The worker, who saw the child on a weekly basis, accepted the mother's self-report that she was administering the primary anti-seizure medication as prescribed and never viewed the medication bottle to determine if there was less after each visit. The worker did contact the child's pediatrician who confirmed that the child was seen regularly and he expressed no concerns for her welfare.

² Although Rett Syndrome is not one of the chronic illnesses discussed in this report, one of the Syndrome's symptoms are seizures. Seizure disorder is covered in the illnesses discussed below.

Two days before her death the worker visited the family for a routine weekly visit. The mother explained that all her children were all home sick with the flu. The child and her siblings had been seen by their pediatrician twice in the past week. The mother reported that while one of her children was still quite ill the child in question was improving. Two days later the child was found unresponsive in the morning after the mother had gone to check on her children. The autopsy, which determined that the child died of complications of Rett Syndrome, also concluded that the child had a therapeutic level of the primary anti-seizure medication in her system at the time of death.

QUESTIONS

1. The case worker was not familiar with Rett Syndrome so she used the internet to become informed about the disorder. The worker was aware that the child had been diagnosed with a seizure disorder and had learned from her research that seizure disorders are common with this disease. While interviewing the mother the worker learned of the mother's belief that her daughter suffered from tremors rather than seizures. What could the worker have done to help the mother better understand the complexities of her daughter's disorder?
2. In this situation, many medical providers were involved in the child's care. Two of the physicians had a difference of opinion regarding the secondary anti-seizure medication. The local pediatric neurologist opined that the medication was necessary and the mother's failure to administer it constituted neglect. The family pediatrician who had consistently cared for the child her entire life felt otherwise. How do you go about reconciling these differences of opinion?
3. The medically complex child was enrolled in special education. Her teacher noted that she had recently been arriving to school in need of a bath. The mother had attempted to obtain a bathing lift through her meagre health insurance but was repeatedly denied. An internet search determined that a bathing chair could have been purchased for somewhere around \$300.00. What could have been done to obtain the bathing chair? Could the case worker have requested Norman funds?

CHAPTER TWO: ASTHMA

OVERVIEW

Asthma is the most common chronic childhood disease; about one in every 10 American children has asthma (U.S. Environmental Protection Agency, 2015b). Asthma is a disease that affects the airways, the tubes that carry air into and out of your lungs. In people with asthma, the airways are inflamed. The inflammation narrows the airway, making it difficult to breathe. The inflammation also makes the airways very sensitive to inhaled substances, like cigarette smoke, pollen, and dust. The airways react to inhaled substances by tightening and producing mucus, which makes it even more difficult to breathe.

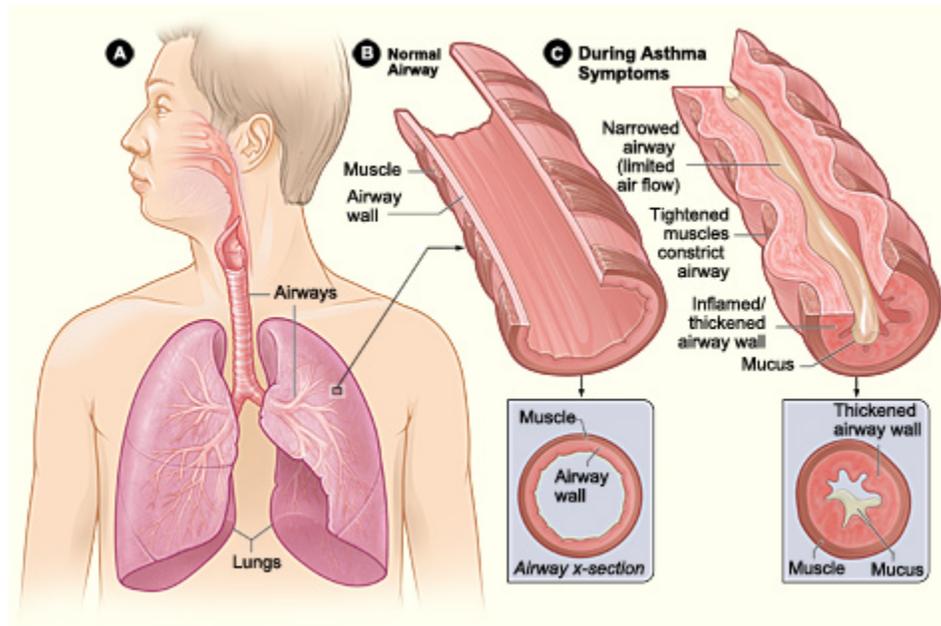


Figure 2.1 Figure A shows the location of the lungs and airways in the body; Figure B shows a cross-section of a normal airway; and Figure C shows a cross-section of an airway during asthma symptoms (National Heart Lung and Blood Institute, 2014).

CAUSE

The exact cause of asthma is unknown. However, researchers believe a combination of genes and environmental factors are likely to play a role. It is thought that individuals are born with specific genes, passed down from parents to children, that put them at risk for developing asthma. Then, exposure to environmental factors that irritate the airways, such as viruses, dust, animal dander, and tobacco smoke, can induce asthma in genetically predisposed individuals.

SIGNS & SYMPTOMS

The signs and symptoms of asthma vary from day to day. Sometimes the symptoms are mild and do not interfere with daily life. Sometimes the symptoms are severe, interfere with daily life, and, possibly, require emergency treatment. The most common signs and symptoms of asthma are coughing, wheezing, chest tightness, and shortness of breath.

COUGHING

Coughing is often worse at night or early in the morning. Sometimes, coughing is so severe that it interferes with sleep.

WHEEZING

Wheezing is a high-pitched whistling sound made while breathing.

CHEST TIGHTNESS

Chest tightness may feel like someone is squeezing your chest or sitting on your chest.

SHORTNESS OF BREATH

Shortness of breath, or dyspnea, may feel like you are out of breath or that you “can’t catch your breath”.

Treat the signs and symptoms of asthma at first notice, as symptoms can quickly become severe. **Severe symptoms can be fatal and require immediate emergency attention.**

DIAGNOSIS

Asthma can occur at any age but most children get symptoms before the age of 5, but it can occur at any time throughout the child’s life, and even into adulthood. One child may have a mild form of asthma, while another may have a form serious enough to threaten the child’s life.

Asthma is a growing problem among children, especially minority children living in crowded cities. One of the difficulties in caring for a child with asthma is realizing it is a chronic disease that needs medication to prevent acute episodes. There is a common myth that children outgrow asthma. This is not true; asthma does not go away. It is extremely important that the child attends regular visits to his/her healthcare provider. The severity of the asthma will determine how often the child needs to be seen. The healthcare team will work with the child’s caretaker to determine what type of asthma the child has, and how best to manage and treat the condition.

TREATMENT

Asthma is a long-term disease that has no cure. Treatment of the disease focuses on preventing and controlling the signs and symptoms. Treatment options differ from person to person, depending on the severity of the disease. Talk to your doctor about what treatment options are best for your child.

AVOID TRIGGERS

Many things can trigger, or worsen, asthma symptoms. The most common triggers are:

- Allergens from dust, animal fur, cockroaches, mold, and pollens from trees, grasses, and flowers
- Irritants, such as cigarette smoke, air pollution, chemicals or dust, and sprays (like hairspray or perfume)
- Sulfites, a preservative found in foods and drinks
- Viral upper respiratory infections, such as colds
- Physical activity, including exercise

While it is not possible to eliminate asthma triggers, there are several things you can do to minimize their effects. These include:

Reduce allergens in the home. Cleaning floors with a wet mop, instead of sweeping, which pushes allergens that have gathered on the floor into the air. Not allowing pets to sleep in the same room with the child. Frequently washing bedclothes and covers to eliminate invisible mold or dust mites. Using allergy proof covers on the mattress and pillows.

Exercise! Even though physical activity can be an asthma trigger, it is an important part of a healthy lifestyle. Do NOT avoid exercise. Medications are available to help ease your asthma symptoms and stay active; talk to your doctor.

Immunizations. Immunizations protect against diseases that could trigger an asthma attack, like the respiratory syncytial virus (RSV) or the influenza virus. People with asthma and all members of the household should be up to date with all childhood vaccines, plus an annual influenza vaccine. People with asthma should ONLY receive the intramuscular, or shot, form of the influenza vaccine; people with asthma should NEVER receive the intranasal form. The intramuscular form is the most effective vaccination against the influenza virus and the intranasal form could trigger an asthma attack.

Working to rid the child's home of triggers may take quite some time and effort, but can be very effective in preventing acute episodes.

MONITOR PEAK FLOW

Peak Expiratory Flow Rate (PEFR), or peak flow, is the maximum speed at which a person can exhale a single breath of air; or, in other words, how well air moves out of your lungs. This can be measured using a small hand held device known as a Peak Flow Meter. Your doctor will show you how to use your peak flow meter and review your technique at each follow-up visit.

Asthma can change gradually. The peak flow can show changes before they can be felt and help predict an asthma attack. It is important to monitor peak flow regularly.

The peak flow meter has three zones: green, yellow, and red.

The **green zone** is 80-100% of your usual peak flow rate. A reading in the green zone means your asthma is under good control and you should continue with your current treatment.

The **yellow zone** is 50-80% of your usual peak flow rate. A reading in the yellow zone means your asthma is getting worse and you may require extra treatment.

The **red zone** is less than 50% of your usual peak flow rate. A reading in the red zone is and **EMERGENCY**. A reading in the red zone means your airways are severely narrow and you need a rescue medication right away. Get medical help **NOW**.

MEDICATIONS

If asthma attacks cannot be prevented by avoiding triggers, medications are available to help control the signs and symptoms.

Rescue medications. These medications work rapidly to relieve the acute symptoms of asthma such as difficulty breathing. They are usually in the form of an inhaler. This group can be used alone only in those who have very mild cases of asthma. These medications are not meant to be used on a regular basis, but rather should be reserved for situations when the child needs immediate relief. The child's healthcare provider will instruct you on how often and how to properly use an inhaler.

Controller medications. These medications reduce the swelling around the air passages in the lung. They are long-term medications that can control asthma by regulating the underlying cause. It is important to know that these medications do not cure asthma but rather keep it under control.

If the healthcare provider finds that the child's asthma is not well controlled with the occasional use of a rescue medication, the provider will prescribe controller medications (inhaler or pills) in order to provide more long-term treatment.

Since these medications act to reduce swelling and work for a longer period of time, the child is less likely to have repeated problems. Some caregivers and parents of children with asthma sometimes mistakenly assume that the child only needs a rescue medication to treat their asthma, when in fact they may need a controller medicine. It is important to discuss the various treatment options with the child's healthcare provider, to determine which medication is the best option. Also, since there can be quite a variety of different types of inhalers, the provider can give you and the child specific instructions for how to use whichever the child needs. You and the child will want to practice using and cleaning the inhaler with the healthcare provider or nurse present, so that you can feel comfortable with its use.

Although the medicines are very important to the child's treatment, be aware that some of these medicines can cause the child to experience undesirable side effects. The rescue medications most commonly cause nervousness, shakiness and poor sleeping. Discuss all possible side effects with the child's doctor. If the child does experience any unexpected side effects, there may be a different medication with fewer problems, which the doctor can prescribe.

PREVENTION

You cannot prevent asthma. However, you can follow the steps mentioned in the treatment section to control the disease and prevent its symptoms.

DOCTOR VISIT

First, when choosing a healthcare provider, you should ask the caseworker to have the child seen by one who has had special training or is familiar in treating childhood asthma. The healthcare provider should develop the Written Action Plan for everyday use, and if the child's asthma is severe, an Emergency Action Plan as well. Ask about how to avoid asthma attacks. You might also meet with a nurse, who can teach you how to do such things as checking the peak flow rate, as well as using and cleaning inhalers. You might also meet with a pulmonologist (lung specialist doctor), who will check the child's lung function. The school nurse will play an important role in helping the child follow the doctor's orders, while in class. The school nurse will likely have additional information about asthma to help the child manage their chronic illness better.

Written Action Plan: The healthcare provider will provide you and your child with a plan for how to care for asthma on an everyday basis. It can contain such information as what type of medication the child should be using (such as inhalers or pills), as well as instructions for what to do if the child's asthma begins to worsen.

Furthermore, the Written Action Plan should also contain information about what to do if you think the medicine is not working and the child is having an asthma attack. Most importantly, it should also describe when it is necessary to go to the hospital.

Emergency Action Plan: It is important to discuss with the child's doctor how to recognize signs that the child is in need of emergency care and/or needs to go to a hospital. Together, you can develop an Emergency Action Plan that will contain this information.

Ask your healthcare provider or caseworker to make multiple copies of both of the action plans (Written Action Plan and Emergency Action Plan) and Asthma Quick Reference Guide so that you always have a copy of each:

- At home
- At the child's school
- With the child whenever they leave the home

Make sure that the Written Action Plan and Emergency Action Plan have the child's name and date so that everyone involved in the child's care can follow the most current plan.

An Asthma Action plan is attached below to bring to your child's doctor's office at their next appointment. It is important you fill this out with your child's healthcare provider. When complete, remember to have the Action plan readily available at your home, at your child's school, and with any caregivers.

The Emergency Action Plan is the basis of keeping the child with asthma safe and preventing an acute episode from turning into a severe asthma attack. It is a good idea to keep copies in convenient places around the home, such as on the refrigerator or next to the telephone. You and the child should review what you would do if the child has an asthma attack that is not getting better after using rescue medicines and following the instructions in the Written Action Plan. Remember that you can always call 911 if you feel that the child is in danger. Although it is very difficult to remain calm in certain situations, it is especially important for the child to feel that you are in control and not leaving him or her while they are in distress. It is also helpful to keep the child as calm as possible.

All caregivers and family members need to be familiar with the acute management of asthma. There are several support groups and camps that children and teens can attend to learn more about asthma and self-care.

Your caseworker can help you obtain information and help get the services that the child needs. Although asthma can be a difficult condition to live with, especially for children, there is a wealth of information available at libraries, the doctor's office, and websites cited at the end of this chapter.

COPING & SUPPORT

Allergy & Asthma Network

Mothers of Asthmatics

www.aanma.org

Environmental Protection Agency

www.epa.gov/asthma

American Lung Association

www.lung.org

Chicago Asthma Consortium

www.cac.com

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CHAPTER THREE: CEREBRAL PALSY

OVERVIEW

Cerebral palsy is a disorder of the brain that affects muscle movement, balance, posture, or motor skills. It is caused by damage to the brain while it is developing, usually before birth. Each child experiences different symptoms depending on where the brain damage takes place. Symptoms may include tremors, eye coordination problems, unsteady gait, stiff or floppy muscles, swallowing problems, seizures, and intellectual disabilities.

CAUSE

Cerebral palsy is caused by damage to the brain that occurs during pregnancy, birth, or right after birth. Usually the exact reason for the damage is not known. However, in some cases it may be related to infection, brain injury, abnormal development of the fetus during pregnancy, premature birth, low birth weight, lack of oxygen during birth, or drug use by the child's mother while pregnant. Cerebral palsy is not contagious. Proper prenatal care can decrease the risk of cerebral palsy; however, it is not possible to fully prevent it.

SIGNS & SYMPTOMS

Every child with cerebral palsy has different symptoms. This is because symptoms depend on where the brain damage occurred. If the brain was injured in the speech area, the child may have difficulty talking and communicating. If the brain was injured in the area that controls urination, they may not be able to get to the bathroom in time to use the toilet. If the brain was injured in the area of the brain that controls chewing and swallowing, they might not be able to safely eat solid foods. Depending on which parts of the brain are affected by cerebral palsy (CP), the child may face some specific health challenges. Some of these challenges include:

BREATHING PROBLEMS

Children with CP are at an increased risk for developing respiratory infections (such as colds and pneumonia). When children with CP have respiratory infections, they can last longer than usual since the child may have difficulty coughing or blowing their nose. In CP, it is also easier for fluids from the nose or from the mouth to get into the lungs. This problem is called aspiration. This can be VERY serious. If you notice that the child has ongoing cough or fever, contact your doctor. If the child has a difficulty breathing, go to the emergency room for evaluation.

MALNUTRITION

Swallowing or eating problems can make it difficult for children who have CP to get enough nutrition. This may cause impaired growth and weaker bones. Children who do not receive enough nutrition may eventually be smaller than their friends and other children their age. Some children may need a feeding tube for adequate nutrition.

- Excessive drooling or problems with swallowing
- Difficulty with sucking or eating

VISION & HEARING PROBLEMS

Partial or total hearing and/or vision loss is possible with CP depending on the part of the brain that is affected. Children should undergo sensory testing to determine if they have limited ability to see or hear, and if they need assistive devices such as glasses or hearing aids. Vision problems are very common in children with CP. Nearly half of all children with CP have an eye muscle problem known as strabismus, commonly known as “cross-eye.” Strabismus causes one of the eyes to turn outward or inward. Sometimes the eyes are unable to focus or track objects. To monitor for this the child’s vision should be checked regularly by your doctor. Some children with CP may have hearing loss. Take note if the child does not blink to loud noises by one month of age, or is not turning his head toward a sound by three to four months, or is not saying words by twelve months. Discuss anything unusual with the child’s doctor.

URINARY AND BOWEL PROBLEMS

Most children with CP are able to be toilet trained. However, it may be a longer process and they may have more trouble either due to developmental delays or due to weak muscles around the bladder. It is important to monitor these children for pain with urination, blood in their urine, fever, and back pain as these may be signs of a urinary tract infection.

Children with CP are at high risk for constipation because their abdominal muscles are weak and they are not as active. It is important for these children to drink lots of fluids, eat fiber, and move as much as possible to prevent constipation. Tracking bowel movements on a calendar can help caretakers to monitor for constipation. Talk to the child’s doctor if the child does not have a bowel movement at least every two to three days, or if their stools are large and hard.

DENTAL PROBLEMS

Children with CP may develop more cavities, because it is hard for them to coordinate their muscles in order to brush their teeth. It is important for the child to get assistance with teeth brushing. Visit a dentist that knows how to care for children with CP.

BONE & JOINT ISSUES

The arms or legs that had been under the control of the part of the brain that was injured may not grow the same way that the other arm or leg grows. If you suspect that there is a difference, it is important to tell the doctor and caseworker. In some cases, a child’s spine may grow curved as well, which is called scoliosis. Depending on the way that the child’s bones grow, they may need a brace or other assistive devices.

People with CP also have higher rates of low bone density (osteopenia) and bone pain and inflammation at joints (osteoarthritis). These issues come from pressure on joints, abnormal joint alignment, lack of mobility, malnutrition, and may be made worse by some anti-seizure medications.

In children with some forms of CP, it is often difficult to prevent contractures. A contracture occurs when muscles tighten unequally, to the point that a child is no longer able to fully move their joint. Contractures can inhibit bone growth, cause bones to bend, and result in joint deformities, dislocation or partial dislocation.

MUSCLE & MOVEMENT ISSUES

CP creates many different muscle problems, depending on what part of the child's brain is affected by CP. Children commonly have variations in muscle tone, including either being too stiff or too floppy. Stiff muscles with exaggerated reflexes are called "spastic." Stiff muscles with normal reflexes are called "rigid." Sometimes children may experience tremors or sudden involuntary muscle movements. Lack of muscle coordination or slow, writhing movements called "athetosis" are also common in children with CP. Because of lack of muscle coordination and weakness these children may struggle to achieve developmental milestones such as walking. Or they may walk on their toes, walk with their knees together, walk with legs far apart, or walk with a crouched gait. Muscle control problems may also lead children to have difficulty with precise movements like picking up a spoon. Finally, often one side of the child's body is more affected than the other, leading to asymmetrical movements.

SEIZURES

Many people with CP also have a seizure disorder. Seizures are brain disturbances in which too many signals happen in the brain at the same time. A seizure can look like uncontrollable full body shaking, muscle twitching, or a blank stare. Please see Chapter Six, starting on page 50, for more information on caring for children with seizures.

COMMUNICATION PROBLEMS

Many children with CP have problems communicating (talking or using sign language), because the muscles of their face and mouth may be weak or hard to control. If a child has a hard time talking, it sometimes makes it more difficult for the healthcare providers and teachers to figure out whether the child has learning problems. Special techniques and therapists may be used with these children to allow them to communicate to the best of their ability.

INTELLECTUAL IMPAIRMENT

Approximately two-thirds of children with CP will also be diagnosed with an intellectual impairment. Intellectual impairment affects a person's ability to communicate, care for self, or socialize. A person with intellectual impairment may have trouble learning, thinking, solving problems, or reasoning. However, since there are several different areas in the brain that can cause cerebral palsy, not all children with CP will have thinking and learning problems or develop mental retardation. It is important not to assume a child with CP has an intellectual impairment until they are diagnosed by a doctor.

DIAGNOSIS

There is no one test for cerebral palsy. To determine if a child has cerebral palsy, doctors will observe the patient, track growth their growth, consult specialists, test blood, and take images of their brain. In severe cases doctors, may see signs of cerebral palsy at birth. However, in most cases it is not identified until between ages 2 and 5 when the child is developing control over their body movements. For this reason, it is important to keep track of a child's progress with developmental milestones like turning from stomach to back and reaching for objects. Ask your case worker about getting an assessment.

MANAGEMENT

Although cerebral palsy cannot be cured, training, therapy, special equipment, and, in some cases, surgery can help a child with cerebral palsy lead a better life. Important goals of treatment for cerebral palsy are to optimize participation in home and community activities.

SPECIAL EQUIPMENT

Your child's doctor or therapist may recommend them using some special equipment to help them get around or do everyday activities. Your child's caseworker or doctor can also give caregivers the information on how to obtain these things. This equipment may include:

Wheelchairs. Wheelchairs are mobile seats that help children who cannot walk get from place to place. Sometimes these wheelchairs can be motorized (the child can "drive" around in their chair) or manual (a child moves the wheels themselves).

Walkers. A walker is a piece of equipment usually made out of light metal that a child places in front of them to help with balance while walking. It most often will have four legs that are adjustable in height and can sometimes have wheels attached.

Adapted silverware and pencils. Children may have a hard time using utensils for eating. Special handles or grips are available for children who have trouble holding onto small objects, such as a fork or spoon.

Communication aids. When a child's speech is very hard to understand, it is easier for them to use a communication aid to talk with others. A communication aid might be a book or poster with pictures that show things the child might want, or an alphabet board that the child can use to spell out their message. There are also computers that are used as communication aids that actually talk for the child.

Helmets. Children may be recommended to wear a helmet to protect their head, in case they fall while walking (remember, balance may be a problem) or have a seizure and fall to the ground. These helmets are similar to those used for bike riding or sports.

DIET

Many children with cerebral palsy have problems growing and developing, because it can be hard to eat and drink. For this reason, there are various techniques for feeding children with cerebral palsy in order to make sure they get all the helpful nutrients they can from their food. If the child cannot be adequately fed by mouth, then the doctor may suggest another way to make sure the child is getting enough calories and fluids.

MEDICATIONS

Oral medications may be used to relax muscles or control seizures in children with cerebral palsy. These medications may sometimes make children drowsy or nauseas. The doctor may also decide to inject medication, such as Botox, to help control involuntary movements. It is important to monitor for any medications side effects that affect the child's ability to interact or perform activities of daily life. If you notice these side effects bring them to the attention of the child's doctor.

SURGERY

Sometimes, surgery may be needed to correct bone or muscle deformities related to cerebral palsy. Surgery may also be used to insert a device that can help control spasticity. Surgery risks and benefits should be discussed in depth with a specialist as surgery is not appropriate for all children.

PREVENTION

Cerebral palsy is not contagious. Proper prenatal care can decrease the risk of cerebral palsy; however, it is not possible to fully prevent it. Most CP develops either during pregnancy or during childbirth, and it is often for unknown reasons. However, with frequent prenatal visits during pregnancy it is possible to decrease preventable causes such as premature birth, drug use during pregnancy, low birth weight, and infection. Sometimes children may acquire CP after birth due to a brain injury through natural (stroke), accidental (car injury) or purposeful (shaken-baby syndrome) means, or if he or she comes down with an infection or a condition that slows blood flow to the brain. For this reason it is important to use an age-appropriate and properly installed car seat, avoid falls, and never shake or hit your child.

DOCTOR VISIT

Children with cerebral palsy require frequent doctor visits. See Navigating a Doctor's Visit (see pages 9 through 12) in the Introduction for information about how to prepare for a doctor visit. Besides the pediatrician, there are other medical professionals who will play an important role in providing health services for the child. Some children with cerebral palsy will see a different medical specialist for each body system, along with physical therapists, occupational therapists, nurses and social workers.

COPING & SUPPORT

Social isolation and the challenges of coping with disabilities can contribute to depression in children with CP. Other children may make fun of the child with CP. This can be very hurtful to a child and limit the child from having friends. The caregivers are very important in listening to a child. If the caregiver thinks the child feels badly, or is developing self-esteem issues, it is important to ask the caseworker and doctor about special help (such as social workers, psychologists, counselors).

Cerebral palsy can be stressful for both people with cerebral palsy and for their care takers. The following organizations offer resources including support groups, education, and referrals:

United Cerebral Palsy of Greater Chicago

Cicero, IL
Phone: 708-863-3803
www.ucpseguin.org

United Cerebral Palsy Of Heartland

Manchester, MO
Phone: 636-227-6030
www.ucpheartland.org

Family Resource Center On Disabilities

Chicago, IL
Phone: 312-9393513
www.frcd.org

United Cerebral Palsy Of Land Of Lincoln

Springfield, IL
Phone: 217-525-6522
www.ucpll.org

If your child has seizures, please also see the Coping & Support section in the Chapter 6: Seizure Disorder (see page 50) for further resources.

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CHAPTER FOUR: CYSTIC FIBROSIS

OVERVIEW

Cystic Fibrosis (CF) is a genetic disorder that affects the mucus glands. In healthy people, the glands produce thin and slippery secretions. In people with CF, the glands produce thick and sticky secretions. The thick, sticky secretions build up, block tubes in the respiratory, digestive, and reproductive systems, and create the perfect environment for bacteria to grow. Over a lifetime, mucus plugs and frequent infections severely damage the organs of the respiratory, digestive, and reproductive tracts.

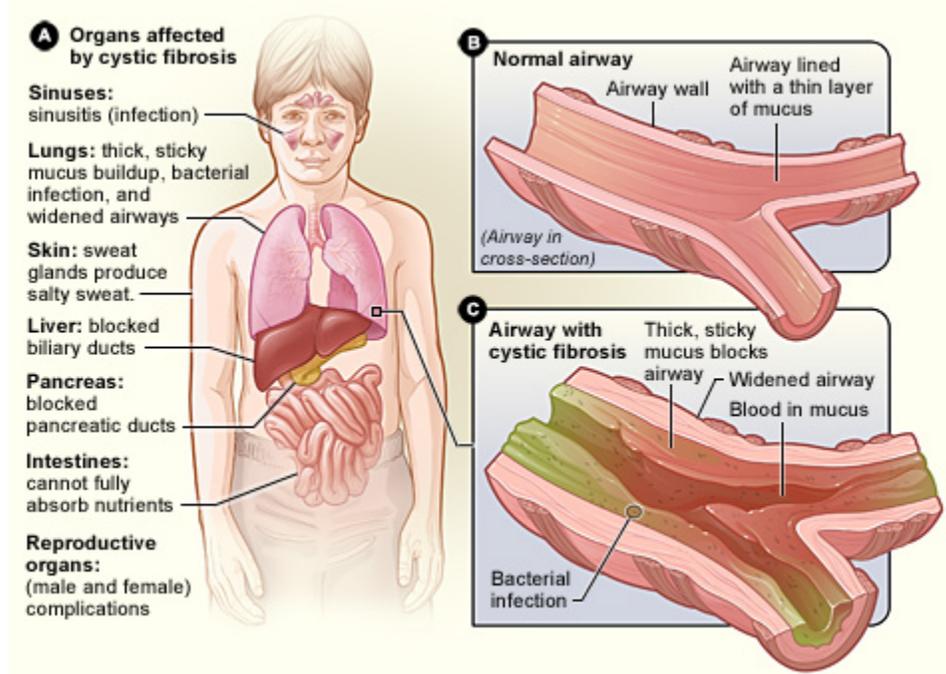


Figure 4.1 Figure A shows the organs that cystic fibrosis can affect. Figure B shows a cross-section of a normal airway. Figure C shows an airway with cystic fibrosis. The widened airway is blocked by thick, sticky mucus that contains blood and bacteria (National Heart Lung and Blood Institute, 2013).

CF is a life-threatening disease. The median predicted life expectancy for someone diagnosed with CF in 2010 is 39 years old (Mackenzie et al., 2014). As treatments for the signs and symptoms of CF continue to improve, so does life expectancy.

CAUSE

People have two copies of most genes, one copy from their father and one copy from their mother. Sometimes mistakes or defects occur in genes and cause genetic disorders. Genetic disorders are passed down in genes from parents to children. They are not contagious and cannot be “caught,” like a cold or flu.

Cystic Fibrosis (CF) is a genetic disorder caused by a defect in the Cystic Fibrosis Transmembrane Regulator gene. It is a recessive disorder, which means a person must inherit two defective copies of the gene to have CF. If only one defective copy of the gene is inherited, the person will be a carrier for CF. CF carriers do not have CF, but can pass the defective gene onto future children. If two normal functioning

copies of the gene are inherited, the person will not have CF and cannot pass the CF onto future children.

The image below shows how the CFTR gene is inherited.

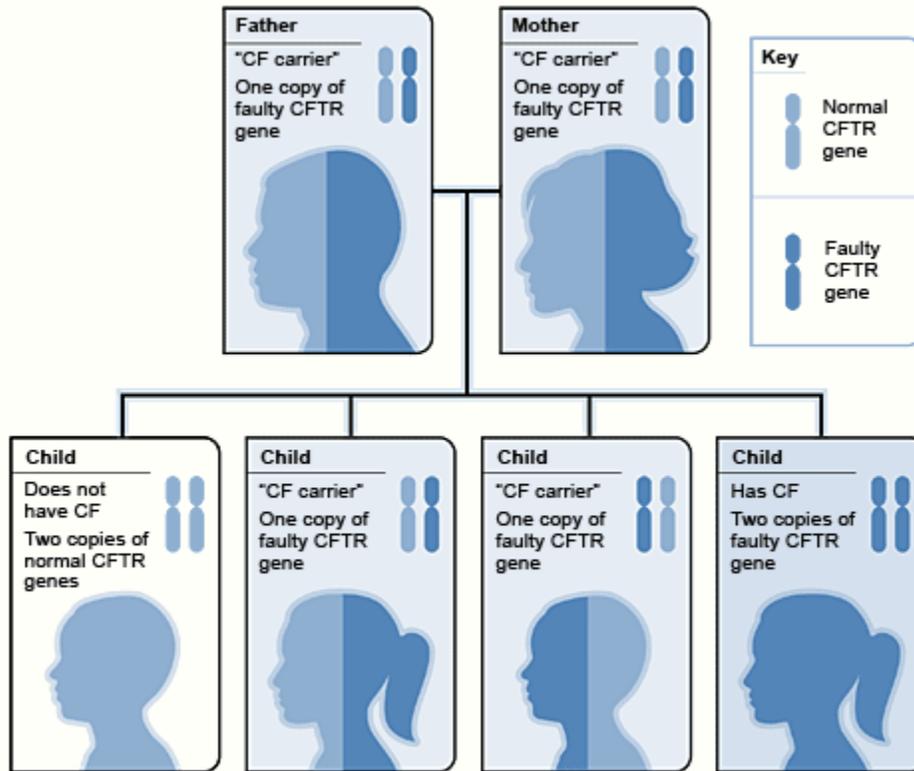


Figure 4.2 The image shows how CFTR genes are inherited. A person inherits two copies of the CFTR gene—one from each parent. If each parent has a normal CFTR gene and a faulty CFTR gene, each child has a 25 percent chance of inheriting two normal genes; a 50 percent chance of inheriting one normal gene and one faulty gene; and a 25 percent chance of inheriting two faulty genes. (National Heart Lung and Blood Institute, 2013).

SIGNS & SYMPTOMS

CF signs and symptoms vary from person to person. Typically, people with CF start to develop signs and symptoms of the disease during the first year of life. Signs and symptoms become more frequent and more severe with age. People with CF are often hospitalized to treat and manage signs and symptoms of CF. The most common signs and symptoms of CF are respiratory problems, frequent infections, digestive problems, reproductive problems, salt loss, clubbing of the fingers and toes, osteoporosis, and nasal polyps.

CF carriers usually do not have any signs or symptoms of the disease and live a normal life.

RESPIRATORY PROBLEMS

Respiratory failure is the most common cause of death in people with CF. People with CF have thick, sticky mucus that builds up in the lungs and blocks the airways. The buildup of mucus makes it difficult to breathe in the necessary amount of oxygen, and oxygen levels in the bloodstream can fall to dangerous levels.

The buildup of mucus also makes it easier for bacteria to grow and causes frequent respiratory infections. Frequent infections damage the airways, making it even harder to breathe.

INFECTION

Cystic Fibrosis does not affect the immune system itself, but people with CF have increased risk for infections. The thick, sticky secretions create the perfect environment for bacteria to grow. Signs of a lung infection include, frequent coughing, bloody sputum, difficulty breathing, chest pain, and fever. Repeated lung infections can cause severe and irreversible damage to the airways.

People with CF also have frequent sinus infections and sinusitis, or inflammation of the sinuses. The sinuses are hollow air spaces around the eyes, nose, and forehead. Signs of a sinus infection or sinusitis are stuffy nose, headache, tenderness under the eyes or at the bridge of the nose, and postnasal drip.

At the first sign of an infection, such as a fever, it is important to see a doctor right away. Early treatment of infection can help prevent problems.

DIGESTIVE PROBLEMS

The pancreas is an important organ of the gastrointestinal and endocrine systems. The pancreas has two major functions. The first function of the pancreas is to produce enzymes that help break down fats and proteins in the small intestines. In people with CF, the thick, sticky mucus blocks the ducts that connect the pancreas to the small intestine and prevents the enzymes from reaching the small intestines. Without the pancreatic enzymes, the small intestines cannot fully digest and absorb fats and proteins. Undigested food causes pain, cramping, gas and either loose, greasy, floating stools or constipation and blockages. In addition, without pancreatic enzymes, nutrients are not absorbed from food and pass through the body without being used. This can cause vitamin deficiencies, malnutrition, and poor growth. It is important for people with CF to visit their primary care doctor on a regular basis to monitor weight and growth rate.

The second function of the pancreas is to produce hormones, such as insulin. Insulin transports sugar from the blood into the cells of the body. Cells need sugar for energy to function. In people with CF, the thick, sticky mucus damages and scars the pancreas. Sometimes, the pancreas is so severely damaged that it can no longer produce enough insulin. This is called cystic fibrosis related diabetes. Children should start being tested for cystic fibrosis related diabetes at 10 years old and should be tested for it every year after that. Cystic fibrosis related diabetes is unique to people with CF, but shares features with both Type 1 and Type 2 diabetes. See Diabetes chapter for more information about diagnosis, signs and symptoms, and treatment of diabetes.

REPRODUCTIVE PROBLEMS

Fertility problems are common in men and women who have CF. Thick, sticky secretions build up in the reproductive tract and make it difficult to conceive. Although CF can cause fertility problems, men and women who have the disease should still have protected sex to avoid sexually transmitted diseases.

SALT LOSS

The Cystic Fibrosis Transmembrane Regulator gene makes a protein that regulates transportation water and salt into and out of cells. In healthy people, the protein reabsorbs the salt in sweat before it reaches the skin surface. In people with CF, the protein does not reabsorb the salt in sweat before it reaches the skin surface. As a result, people with CF lose more salt than normal in their sweat. Losing too much salt is dangerous. Signs and symptoms of losing too much salt are tiredness, weakness, vomiting, muscle

cramps, and muscle aches. If you suspect your child has lost too much salt, contact your primary care doctor.

It's recommended for people with CF to drink lots of fluids, especially during the summer months when children are active and tend to sweat more. It is also recommended that children with CF eat a high-salt diet to replace salt lost in sweat.

CLUBBING OF FINGERS AND TOES

Clubbing is the widening and rounding of the tips of your fingers and toes. Clubbing occurs when there is not enough oxygen in the bloodstream.

OSTEOPOROSIS

Healthy bones require vitamin D. Vitamin D is fat-soluble, which means it needs fat to be absorbed. Since people with CF have trouble digesting fat, they usually have trouble absorbing vitamin D, as well. Without enough vitamin D, their bones can become weak and brittle, also known as osteoporosis.

NASAL POLYPS

People with CF tend to have nasal polyps, or fleshy growths, inside their noses. It is not clear how CF causes polyps. Nasal polyps can grow large enough to obstruct airflow. If polyps start to cause complications, they can be surgically removed.

DIAGNOSIS

CF can be diagnosed before birth with two prenatal genetic tests, amniocentesis and chorionic villus (CVS). In amniocentesis, a doctor inserts a hollow needle through the abdominal wall into the uterus and removes a small amount of fluid from the sac around the baby. The fluid is tested to see whether both of the baby's genes are normal. In CVS, a doctor threads a thin tube through the vagina and cervix to the placenta and removes a tissue sample from the placenta using gentle suction. The tissue is tested to see whether both of the baby's genes are normal.

CF is diagnosed after birth with a blood test, genetic test, or sweat test. The blood test measures pancreatic enzymes and determines whether a newborn's pancreas is working properly. The genetic test shows whether a newborn has defective genes. If a genetic test or blood test suggests CF, a doctor will confirm the diagnosis using a sweat test. For this test, the doctor triggers sweating on a small patch of skin on an arm or leg. Sweat is collected on a pad of paper and then analyzed. High levels of salt in sweat confirm a diagnosis of CF.

Early diagnosis of CF is important. In the United States, all states are required to test for CF as part of the newborn screening. It is recommended that caseworkers and caregivers obtain a copy of the newborn screening to keep with the child's home medical record.

MANAGEMENT

Currently, there is no cure for CF. Management of the disease is largely developed around preventing complications and treating the signs and symptoms of CF. As treatments for the signs and symptoms of CF continue to improve, so does life expectancy for those who have the disease. Today, some people with CF are living into their forties or fifties.

RESPIRATORY COMPLICATION

There are many ways that caregivers can prevent or reduce the number of respiratory complications.

ELIMINATE TOBACCO SMOKE

Research has shown that just being around tobacco smoke can be harmful to children with CF. Eliminating exposure to tobacco smoke can help improve the health and lung function of children with CF.

EXERCISE

Caregivers should encourage children with CF to exercise and participate in games/physical activities as much as they are physically able. Exercise helps to loosen mucus buildup in the lungs and can strengthen the lungs. Children with CF may not have as much energy as other children their age, so it is important to talk to your primary care doctor to determine the appropriate level of exercise.

COUGH

People with CF will frequently cough to clear the mucus that clogs the lungs and airways. Caregivers should encourage children to cough because it helps them breathe easier. Caregivers should not try to stop the child from coughing by giving cough medicines or suppressants.

AVOID ALLERGENS

Exposure to allergens can cause the muscles of the respiratory system to contract, narrowing the airways, and making it difficult to breathe. Eliminating dust, carpeting, fur, feathers, stuffed animals, and pets can also help reduce the number of allergens in a home. Special air purifiers with HEPA filters can help reduce the number of allergens in a home.

CARDIO PULMONARY RESUSCITATION (CPR) CERTIFICATION

CPR is an important technique that caregivers should learn in case of emergency. CPR should only be performed when the child is not breathing and does not have a pulse. Contact your caseworker to help you find a training course near your home.

CHEST PHYSICAL THERAPY (CPT)

People with CF need a way to physically remove thick mucus from their lungs. This is often done by manually clapping on the front and back of the child's chest to help dislodge mucus from the airways or with the help of devices, like high-frequency chest wall oscillation or positive expiratory pressure masks. A primary care provider can teach caregivers how to properly administer CPT.

MEDICATIONS

Antibiotics. Antibiotics are medications that are used to treat or prevent bacterial infections. Antibiotics can be swallowed, inhaled, or administered directly into a vein (intravenous). Oral antibiotics often are used to treat mild lung infections. Inhaled antibiotics are often used to prevent infections. Intravenous antibiotics are used to treat severe or hard-to-treat infections and are given in a hospital.

Corticosteroids. Corticosteroids are medications that reduce swelling in the airways. Corticosteroids can be swallowed or inhaled.

Bronchodilators. Bronchodilators are medications that relax the muscles of the airways. Bronchodilators are inhaled. They are often taken just before CPT to help clear mucus out of the child's airway or just before inhaling other medications to increase delivery of medication to the lungs.

Mucolytics. Mucolytics are medications that breakdown mucus, making mucus thinner and easier to expel. Mucolytics are inhaled.

OXYGEN

Eventually, the lungs will become severely damaged by mucus plugs and frequent lung infections. When this occurs, the lungs will no longer be able to inhale the necessary amount of oxygen and oxygen levels in the bloodstream will decrease to dangerous levels. It may become necessary to begin oxygen therapy to increase oxygen levels in the bloodstream.

LUNG TRANSPLANT

If all other treatments do not work and lung function continues to decline, a lung transplant is the last available option. A lung transplant is surgery to remove a person's diseased lung and replace it with a healthy lung from a deceased donor.

INFECTION

There are ways that caregivers can help prevent or reduce the number of infections:

Handwashing. Handwashing is the number one way to prevent the spread of germs.

Antibiotics. Antibiotics kill or stop the growth of bacteria and are also used to treat bacterial infections.

Immunizations. All members of the household should be up to date with all recommended childhood vaccines, plus influenza, pneumococcal, and meningococcal vaccines.

DIGESTIVE PROBLEMS

People with CF require a special diet to digest food and absorb nutrients. Children with CF require high-calorie, high-protein, high-fluid diets supplemented with enzymes, salt, and vitamins. It is important to discuss dietary needs with a doctor or nutritionist. Below is a list of commonly given supplements to aid with digestion:

Enzymes. Children with CF need to take pancreatic enzyme supplements with each meal. Pancreatic enzymes help the body digest and absorb nutrients from food. The enzyme dose depends on the type and amount of food eaten. A doctor and nutritionist will help you understand the appropriate amount of enzymes to give the child. Enzymes should be taken 30 minutes before eating, and the beads of the capsule should not be chewed or crushed.

Salt. Since a large amount of salt is lost in sweat, salt and foods high in salt content are recommended for all ages to replace the amount lost. It's recommended for people with CF to drink lots of fluids, especially during the summer months when children are active and tend to sweat more. It is also recommended that children with CF eat a high-salt diet to replace salt lost in sweat.

Vitamins. Vitamins may be poorly absorbed because of incomplete digestion of fats. For this reason, it is recommended that a child with CF take a daily multivitamin.

Fluids. Encourage your child to drink plenty of fluids. This is especially important during the summer months, when children are active and tend to lose a larger amount of water in sweat.

PREVENTION

You cannot prevent cystic fibrosis. However, you can follow the steps mentioned in the management section to control the disease and prevent its symptoms.

DOCTOR VISIT

Children with CF require frequent doctor visits. See *Navigating a Doctor's Visit* (pages 9 through 12) in the Introduction for information about how to prepare for a doctor visit.

Care plans will vary from child to child. Some children will need to visit the doctor more often than others. In addition to routine primary care doctor visits, children with CF should also visit a CF specialist, genetics counselor, and vision/hearing screenings.

CF SPECIALIST – EVERY 3 MONTHS

The CF Foundation recommends children with CF visit a CF specialist every 3 months. At the visits, the doctor will monitor physical growth and development and respiratory function.

To find a CF specialist located near you, visit the CF Foundation website at www.cff.org. There are over 100 CF care centers in the US and a complete list of CF care centers can be found on the CF Foundation website.

GENETICS COUNSELOR – AT PUBERTY

It is recommended that children with CF or CF carriers receive genetics counseling when they reach puberty or sexual maturity. Children with CF or CF carriers can pass the gene for CF onto their children. A genetics counselor will perform genetic testing to determine if they are at risk for having a child with CF. A genetics counselor can also answer any questions and explain the choices that are available. If you do not already have a genetics counselor, contact your primary care doctor or CF specialist for a referral.

VISION & HEARING – EVERY YEAR

Vision and hearing screenings should be performed annually. CFRD can damage the blood vessels in the eye and cause blindness. A common group of antibiotics used to treat infections related to CF, called aminoglycosides, can cause damage to the structures of the ear.

COPING & SUPPORT

It is important to know that chronic health conditions can have a powerful effect on how children think about themselves and their ability to relate to others. Encouraging children to talk about how they feel about themselves and how they think others see them can help, both emotionally and socially. If the caregiver feels the child has low self-esteem, professional help can be sought. Caseworkers, doctors, and other health care professionals can work to uncover underlying issues that are causing low self-esteem. Below are some underlying issues that may prevent a child with CF from feeling good about him or herself.

- Children with Cystic Fibrosis are often shorter, have difficulty gaining weight, and experience delayed sexual maturity/puberty. These differences may lead to people staring at, making fun of, or excluding your child from activities.
- Sometimes, people think CF is contagious and children won't play with your child; or, if it is an adult, let your child play with their children. This reaction towards your child is the result of their fear and lack of knowledge about CF.
- Children with CF might not be able to fully participate in some activities, such as sports, and feel left out.
- Children with CF may be teased or picked on at school because of their frequent cough.
- Taking tablets or capsules with their meals may be embarrassing for them.
- Bowel movements of children with CF typically have a strong, bad odor. This might be embarrassing for the child and they may require extra privacy when toileting.

In addition, community centers, CF clinics, and CF organizations are great sources of support and information for both the child and caregiver. Below are some helpful CF organizations:

Cystic Fibrosis Foundation, USA

www.cff.org

Cystic Fibrosis Trust, UK

www.cysticfibrosis.org.uk

Cystic Fibrosis Worldwide

www.cfww.org

Cystic Fibrosis

www.cysticfibrosis.com

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CHAPTER FIVE: DIABETES, TYPE 1 AND TYPE 2

OVERVIEW

Diabetes is a disease characterized by high blood sugar, also called blood glucose. Glucose is the body's main source of energy and comes from carbohydrates that you eat. In healthy people, insulin, a hormone produced by the pancreas, transports glucose from the blood into the cells of the body to be used as fuel. In diabetes, the body doesn't make enough insulin, doesn't use insulin well, or both. Thus, glucose cannot enter the cells and the cells starve. In addition, glucose builds up in the blood and damages organs throughout the body, such as the eyes, skin, and kidneys.

There are many different types of diabetes. The three most common are type 1, type 2, and gestational diabetes. This chapter will focus only on type 1 and type 2 diabetes.

TYPE 1 DIABETES

CAUSE

Currently, there is no known cause for type 1 diabetes. Type 1 diabetes is characterized by autoimmune destruction of the beta cells of the pancreas. This means the body's own immune system attacks and destroys the cells that make insulin. Once the beta cells are destroyed, the pancreas can no longer produce insulin. So, people with type 1 diabetes do not make enough, if any, insulin and must take insulin every day to live.

SIGNS & SYMPTOMS

Diabetes signs and symptoms vary depending on your blood glucose level, whether it is high or low. High and low blood sugar can be equally dangerous.

HYPERGLYCEMIA

A blood glucose level greater than 240 mg/dl is considered HIGH blood glucose, also called hyperglycemia. Hyperglycemia can have many causes. Eating more food than planned or foods that are high in carbohydrates, exercising less than planned, and missing insulin doses or not taking enough insulin are the most common causes of hyperglycemia. Illness, growth spurts, stress, and certain medications can also induce hyperglycemia.

The primary signs and symptoms of hyperglycemia are polyphagia (eating a lot), weight loss, polydipsia (drinking a lot), and polyuria (urinating a lot).

POLYPHAGIA (INCREASED HUNGER)

When there is a deficiency of insulin, glucose cannot enter the cells and the cells start to starve. The brain recognizes that the cells are starving and stimulates the body to eat.

WEIGHT LOSS

When there is a deficiency of insulin, the body breaks down fat to use for energy. As fat stores are depleted, you start to lose weight.

POLYURIA (INCREASED URINATION)

Polyuria, or increased urination, is the kidneys attempt to eliminate excess blood glucose in urine.

POLYDIPSIA (INCREASED THIRST)

Polydipsia, or increased thirst, is the body's attempt to compensate for increased urine output.

DIABETIC KETOACIDOSIS (DKA)

DKA is a medical emergency and should be treated in a hospital. Seek emergency care if blood sugar is higher than 300 mg/dl and you are exhibiting signs and symptoms of DKA. If left untreated, DKA can be fatal.

All cells in the body need to use glucose for fuel to survive. Without enough insulin, the glucose cannot enter your cells and your body begins to break down fat to use as fuel instead. The breakdown of fat creates high levels of blood acids, called ketones, and leads to diabetic ketoacidosis (DKA).

Signs and symptoms of DKA include:

- Excessive thirst
- Frequent urination
- Nausea and vomiting
- Abdominal pain
- Weakness or fatigue
- Loss of consciousness
- Confusion
- Shortness of breath
- Fruity-scented breath

HYPOGLYCEMIA

A blood glucose level less than 70 mg/dl is considered LOW blood glucose, also called hypoglycemia. All children who take insulin are at risk for hypoglycemia. Hypoglycemia is one of the most immediate emergencies for a person with diabetes, because the signs and develop very quickly without warning (Taliaferro, 2013a). Hypoglycemia can occur when exercising more often, taking too much insulin, eating less food or skipping meals.

The primary signs and symptoms of hypoglycemia are shakiness and jitteriness, sweating, paleness, hunger, blurry vision, confusion or disorientation, and being weak or lethargic. Children with diabetes should be educated to keep a supply of fast-acting carbohydrates with them always, to treat hypoglycemia at the first sign. Examples of fast acting carbohydrates include: fruit juice (not low-calorie or reduced-sugar), regular soda (not low-calorie or reduced-sugar), and glucose tablets or gel. Loss of consciousness, seizures, and inability to swallow can occur when hypoglycemia progresses to severely low blood glucose levels. Severely low blood glucose levels is a medical emergency and should be treated in a hospital.

DIAGNOSIS

Type 1 diabetes can occur at any age, but is typically diagnosed in children and young adults (NDEP, 2016). To diagnose diabetes, and differentiate type 1 diabetes from type 2 diabetes, your PCP will perform a variety of tests:

GLYCATED HEMOGLOBIN (A1C) TEST

This blood test shows your average blood sugar level for the past two to three months. It measures the percentage of blood sugar attached to the oxygen-carrying protein in red blood cells, called hemoglobin. The higher your blood sugar levels, the more sugar you'll have attached to hemoglobin. An A1C level of 6.5% or higher on two separate tests indicates diabetes. A normal A1C level is less than 5.7%.

RANDOM BLOOD SUGAR TEST

A blood sample will be taken at a random time. Regardless of when you last ate, a random blood sugar level of 200 mg/dL or higher suggests diabetes.

C-PEPTIDE TEST

C-peptide is a byproduct of insulin production. C-peptide is a sign that your body is producing insulin. A low level, or no C-peptide, indicates that your pancreas is producing little or no insulin, which is indicative of type 1 diabetes.

MANAGEMENT

Everyone with diabetes should have a diabetes management plan, developed by a primary care provider and diabetes care team. Diabetes management plans are individualized and will be different for each person. The goal of all diabetes management plans is to balance a child's diet, exercise, and medication needs.

MONITOR BLOOD GLUCOSE

Blood glucose levels change in response to food, exercise, alcohol, illness, and medications. Blood glucose should be carefully monitored before each meal, before and after exercise, and before bed. The easiest way to monitor your child's blood glucose is with a home glucose meter or glucometer. A nurse or other healthcare professional will teach you and your child how to use a glucometer at home.

Target blood glucose levels are individually determined by your primary care doctor and diabetes care team. For many children who have diabetes, the below blood glucose values are generally recommended:

Under 5 years:	80-200 mg/dl
5-11 years:	70-180 mg/dl
12 years and up:	70-150 mg/dl

MEDICATION

Everyone with type 1 diabetes must take insulin daily. Insulin can be administered via needle and syringe, insulin pen, insulin jet injector, and/or insulin pump. There are four types of insulin: rapid-acting, short-acting, intermediate-acting, and long-acting. Each type of insulin has a different onset (starting time) and duration (how long it lasts in the body).

Type of Insulin	Medication Name Brand (Generic)	Onset	Duration
Rapid-acting	Humalog (lispro) NovoLog (aspart) Apidra (glulisine)	15 minutes	3-5 hours
Short-acting	Humulin R Novolin R	30-60 minutes	5-8 hours
Intermediate-acting	Humulin N Novolin N	1-3 hours	12-16 hours
Long-acting	Lantus (glargine) Levemir (detemir)	1 hour	20-26 hours

Your doctor will determine what type of insulin should be used and the best way to administer it. Often, your child will be on more than one type of insulin to maintain stable blood glucose levels throughout the day.

DIET

Eating healthy is an important part of managing diabetes. Your doctor will likely recommend that you see a dietitian to develop a healthy eating plan. The plan will help your child keep blood glucose levels in a safe range by making healthy food choices. It is important for your child to follow meal plans your nutrition team provides you.

A diabetes diet is based on eating three meals a day at regular times, limiting foods that are high in sugar, eating smaller portions, monitoring carbohydrate intake, and limiting fat, salt, and alcohol intake.

RECOMMENDED FOODS

Healthy (or Complex) carbohydrates. During digestion, sugars (simple carbohydrates) and starches (complex carbohydrates) are broken down into glucose. Simple carbohydrates are found in “junk food”, such as candy and soda pop, and offer little nutritional value. Complex carbohydrates are healthy and keep your body fueled all day long. Complex carbohydrates can be found in whole grains, fruits, vegetables, beans, and dairy products. In addition to choosing healthy carbohydrates, people with type 1 diabetes also need to track their carbohydrate intake, to make sure they are not getting too much or too little. There are two ways to track carbohydrate intake. Your child can consume the same amount of carbohydrates with each meal, and, then, take the same dose of insulin at the same time every day. Or, your child can count the carbohydrates eaten with each meal and adjust the amount of insulin given.

Fiber-rich foods. Dietary fiber includes all parts of plant foods that your body can't digest or absorb. Fiber moderates how your body digests and helps control blood sugar levels. Foods high in fiber include vegetables, fruits, nuts, legumes (beans, peas and lentils), whole-wheat flour and wheat bran.

Heart-healthy fish. Fish can be a good alternative to high-fat meats. For example, cod, tuna and halibut have less total fat, saturated fat and cholesterol than beef and poultry. Fish such as salmon, mackerel,

tuna, sardines and bluefish are rich in omega-3 fatty acids, which promote heart health by lowering blood fats called triglycerides. Avoid fried fish and fish with high levels of mercury, such as tilefish, swordfish and king mackerel.

"Good" fats. Foods containing unsaturated fats are healthy in moderation and can help lower your cholesterol. Unsaturated fats can be found in avocados, almonds, pecans, walnuts, olives, and canola, olive and peanut oils. Don't overdo it, though, as all fats are high in calories!

FOODS TO AVOID

Unhealthy carbohydrates. It is important to limit, but not eliminate simple carbohydrates, such as cookies, ice cream, and cake. Plan for occasional sweets, so that your child does not feel like they are being "punished" because of their diabetes. Your dietician can let you know when the best time of day is to allow sugary snacks, so that blood glucose levels stay within control.

Saturated fats. Eating foods high in saturated fats raises your cholesterol and increases your risk of heart disease and stroke. Saturated fats are typically solid at room temperature, such as butter, High-fat dairy products, and animal proteins such as beef, hot dogs, sausage and bacon.

Trans fats. These types of fats are found in processed snacks, baked goods, shortening and stick margarines. Avoid these items.

Cholesterol. Sources of cholesterol include high-fat dairy products and high-fat animal proteins, egg yolks, liver, and other organ meats. Aim for no more than 200 milligrams (mg) of cholesterol a day.

Sodium. Aim for less than 2,300 mg of sodium a day. However, if you also have hypertension, you should aim for less than 1,500 mg of sodium a day.

EXERCISE

Everyone needs regular exercise, including people with diabetes! Encourage your child to participate in physical activities that he/she enjoys, like biking, swimming, or playing tag. Aim for at least 30 minutes of physical activity five days of the week. Remember, physical activity lowers blood glucose. Make sure to measure blood glucose before and after any activity. Your child might need to eat a snack before or after the activity to make sure the blood glucose does not drop too low. Ask your caseworker for community resources such as your local YMCA for activities for your child.

PREVENTION

Currently, you cannot prevent your child from getting type 1 diabetes. Type 1 diabetes is not contagious and cannot be "caught", like a cold or flu. Eating too much sugar also does not cause type 1 diabetes.

TYPE 2 DIABETES

CAUSE

Most children who are diagnosed with type 2 diabetes are overweight or obese. High-fat diets, large portion sizes, and inactivity are causing more children to be diagnosed with type 2 diabetes. In type 2 diabetes the pancreas can make insulin but the body does not use the insulin properly. This is called insulin resistance. At first the pancreas tries to make extra insulin to make up for the insulin resistance but overtime the body cannot keep up and make enough insulin to keep blood glucose at normal levels.

Type 2 diabetes can be prevented by maintaining a healthy lifestyle. Children can manage type 2 diabetes with lifestyle changes, insulin therapy, and oral medications.

SIGNS & SYMPTOMS

Type 1 and type 2 diabetes have the same signs and symptoms. (See pages 40 and 41.)

DIAGNOSIS

Type 2 diabetes can be diagnosed at any age. Many of the same tests used to diagnose type 1 diabetes will be used to diagnose type 2 diabetes. Two additional tests that may be performed are:

Oral glucose tolerance test (OGTT). This is a 2-hour test that checks your child's blood glucose before and 2 hours after your child drinks a special sweet drink. After 2 hours, if blood glucose is equal to or greater than 200mg/dL, a diabetes diagnosis will be given. A normal OGTT is less than 140 mg/dl.

Fasting plasma glucose (FPG). This test checks your child's fasting blood glucose levels. Fasting means not eating or drinking in the last 8 hours. The test is usually done in the morning before breakfast after your child has slept. Diabetes is diagnosed when fasting blood glucose is 126mg/dl or higher. A normal FGP is less than 100 mg/dl.

MANAGEMENT

Some people with type 2 diabetes can achieve their target blood glucose levels with diet and exercise alone. Some people, on the other hand, also need diabetes medications or insulin therapy to achieve their target blood glucose levels.

MONITOR BLOOD GLUCOSE

If your child requires insulin to manage type 2 diabetes, your doctor may recommend daily blood glucose testing. How often you will need to monitor your blood glucose depends on the type and amount of insulin your child needs. Testing is usually recommended before meals and at bedtime. If your child manages type 2 diabetes with noninsulin medications or with diet and exercise alone, you may not need to test your blood sugar daily; consult your doctor.

DIET & EXERCISE

Diet and exercise are just as important in the management of type 2 diabetes, as in type 1 diabetes. Eating a healthy diet of mostly complex carbohydrates, fiber rich foods, lean protein, and good fats can help control your child's blood glucose levels. Additionally, aim for your child to have at least 30 minutes of physical activity five days a week to maintain a healthy lifestyle and help control his/her type 2 diabetes. See Diet and Exercise in the Type 1 Diabetes section (see pages 43 and 44) for additional healthy eating and exercise tips.

MEDICATIONS

Some people with type 2 diabetes do not need medications, and can achieve their target blood glucose levels with diet and exercise alone. On the other hand, some people need medications to achieve their target blood glucose levels. There are many different medications that help control blood glucose. Your doctor will determine which medication is right for your child and describe how each medication works.

PREVENTION

Unlike type 1 diabetes, type 2 diabetes is preventable! Unhealthy eating habits and inactivity are the main factors that put your child at risk for type 2 diabetes. To prevent type 2 diabetes, encourage your child to eat a HEALTHY DIET, rich in complex carbohydrates, lean proteins, and low-fat dairy products, and to limit simple carbohydrates, fat, salt, and alcohol intake. Also, encourage your child to be ACTIVE! Staying active, playing sports, and limiting screen time (i.e. videogames and television) can reduce weight gain and help prevent type 2 diabetes. Ask your case manager for assistance in locating resources for activities.

DOCTOR VISIT

During each healthcare visit, the doctor will often check several things. The physician will monitor if glucose or ketones are in the urine (ketoacidosis), and checks the child's eyes, skin, and sensation for possible damage. The doctor will want to know if the child's current type of insulin treatment is working. A special test called A1C can be performed to see how well the insulin and diet have been working over the last 6-8 weeks. The A1C checks how much glucose is sticking to the oxygen carrying part (hemoglobin) of the red blood cell.

Children who take insulin shots are often recommended to have A1C performed four times a year. Children who do not take shots may only need to have this test performed twice per year. It is a good idea to discuss the results with the doctor and keep track of what he or she says by writing it down in a log or chart.

COPING & SUPPORT

Children are sometimes embarrassed or angry because they have diabetes. They think that their diabetes will prevent them from playing with others or leading a normal childhood life. As with any chronic disease, sometimes children with diabetes may become sad or even depressed. It is important to talk to your doctor about possible signs of depression in the child, because depression could also indicate that the sugar level of the child is not where it should be (too high or too low). If the sugar level is out of control, it can cause the child to act differently.

For example, high or low blood sugar levels during the day might make the child feel tired or anxious. In addition, low blood sugar levels can sometimes lead to hunger and overeating. At night, low blood sugar levels can make it difficult to sleep. However, these symptoms are also common signs of depression. That is why it is important to note differences in the child and tell the doctor. Checking the child's levels of blood sugar may help to explain changes in the child's mood or behavior.

How does a child with Diabetes relate to others and manage stress?

Children with diabetes can sometimes experience fluctuations in blood glucose levels due to increased amounts of stress. Increased stress can develop from being sick, participating in highly physical activity, or even emotional events (such as issues with family, school or friends). Stress affects everyone differently, but there are a few things to consider with children. First, children under stress may not take care of themselves. They may start eating junk food and exercising less, forget to check their glucose levels, or forget to take insulin shots. Secondly, stress hormones (proteins secreted by the body) may alter blood glucose levels directly. If not monitored carefully, physical and mental stress can lead to dangerously drastic changes in blood glucose levels.

Things to help children with diabetes at school:

- Inform the school principal, teacher and nurse that the child has diabetes.
- Provide emergency contact information.
- Provide specific information about the child's diabetes (such as the type and amount of insulin they are taking).
- Provide the school with all necessary equipment for your child's diabetic medical care (such as glucose test kit, insulin shot/injector/pump, sheet to write down glucose levels, etc.) and Individual Education Plan or Section 504 Plan or other education plan.
- Inform school staff of any changes in child's health.
- A bracelet is a good idea for the child to wear that says that the child has diabetes. Many bracelets are available for children and teens and they are designed especially for young people. You can ask the nurse about them.
- Discuss with the child the importance of eating meals that are specially designed for their diabetes.
Make sure that the child has emergency snacks, such as crackers, with them at all times in case their blood glucose gets too low.

Many challenges come with caring for a child with diabetes. For children, these challenges come from eating the right foods, drinking the right drinks, monitoring their glucose and taking their insulin injections or pills. It can seem to be a lot for anyone, let alone a child. It is important to try not to be discouraging to the child by discussing their glucose numbers as "bad." Instead, try to tell the child that the numbers will change and the goal is to stay within the optimal range. Over time, caregivers find it easier to make meals with the proper nutrient source for a child with diabetes. Another challenge caregivers may face is monitoring the child's glucose levels and administering insulin shots. Trying creative ways to chart the glucose testing can make this seem less upsetting. It is never easy to be "stuck" in order to get the drop of blood needed for monitoring. Children (and adults) can get very tired of being "stuck" and sometimes will want to quit. Be prepared to deal with the child who refuses to have their glucose levels read or to take their insulin shots. Try to be sympathetic to a child with diabetes. Sometimes they may feel like a "pin cushion" or as though the caregiver pays more attention to their diabetes than to them. Offer them comfort and support when they need it and encourage them to play a role in their own health care needs.

American Diabetes Association

1701 N Beauregard Street
Alexandria, VA 22311
Phone: 800-342-2383
www.diabetes.org

Academy of Nutrition and Dietetics

120 S Riverside Plaza, Suite 2000
Chicago, IL 60606
Phone: 800-877-1600
www.eatright.org

Centers for Disease Control and Prevention

1600 Clifton Road
Atlanta, GA 30333
Phone: 800-232-6348
www.cdc.gov/diabetes

Joslin Diabetes Center

1 Joslin Center
Boston, MA 02215
Phone: 888-JOSLIN-2
www.joslin.harvard.edu

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CHAPTER SIX: SEIZURE DISORDER

OVERVIEW

Epilepsy is a neurological condition that affects the nervous system. Epilepsy, also known as a seizure disorder, causes a person to have seizures. A seizure is a disruption in communication between cells in the brain. When a person has a seizure, they may experience uncontrollable full body shaking, fainting, twitching of specific muscles, tremors, or a blank stare that looks like daydreaming. How a person's body looks during a seizure depends on the part of their brain where the signal disturbance is taking place. Generally, seizures last for 1-2 minutes. After a seizure a child is often disoriented or very sleepy.

CAUSE

A seizure is a disturbance of signals in the brain. Billions of nerve cells exist in the brain. These nerve cells transmit signals and stimulation to and from the rest of the body. A person has a seizure when too many nerves cells stimulate the brain at one time. This can be caused by either a problem in the brain or a problem in the body.

SIGNS & SYMPTOMS

Seizures are separated into different categories based on where in the brain the seizure begins, the parts of the brain are affected, and what movements and parts of the body are involved. There are three major groups of seizures: generalized onset, focal onset, and unknown onset.

GENERALIZED ONSET SEIZURES

Generalized onset seizures start on both sides of the brain at the same time. The signs and symptoms of generalized onset seizures vary depending on the seizure type.

ABSENCE

- Blank stare, eye-blinking or mouth movements for 10-20 seconds at a time
- Often occurs without being noticed
- Most common in children 4-14 years old

ATONIC

- Total loss of muscle tone and consciousness.
- Child falls to the ground during the seizure, placing them at risk for head injury or other serious trauma.
- Also called "drop" seizures

MYOCLONIC

- One or a series of brief, shock-like jerks of a muscle or group of muscles
- Because seizure is so brief, child is usually awake and able to think clearly.
- May be mistaken for tics, tremors, or clumsiness

TONIC-CLONIC

- Loss of consciousness and body stiffening (tonic stage), followed by shaking of the arms, legs, and face (clonic stage)
- After the seizure stops, generally there is a recovery period in which the child is confused
- The most common type of generalized seizure

STATUS EPILEPTICUS

- When a seizure lasts more than 5 minutes, or child does not regain consciousness between seizures
- This is a medical emergency
- Can result in brain damage or death in the most severe cases.
- If your doctor prescribed a medication to be given at home during seizures, give it now
- **Call 911!**

FOCAL ONSET SEIZURES

Focal onset seizures, previously known as partial seizures, start on one side of the brain. The signs and symptoms of focal onset seizures vary depending on the seizure type.

FOCAL ONSET AWARE (PREVIOUSLY KNOWN AS SIMPLE-PARTIAL)

- Occur when a person is awake and aware during a seizure
- Usually very short, and may include sudden jerking movements or abnormal sensory issues (sight, smell, hearing, touch, taste, hallucinations).
- Does not affect the child's consciousness or memory
- There may be a short period of weakness or loss of sensation after the seizure has ended.

FOCAL ONSET IMPAIRED (PREVIOUSLY KNOWN AS COMPLEX-PARTIAL)

- Occurs when a person is confused or their awareness is affected in some way during a seizure
- During these seizures a person is generally semi-conscious. They may look awake, but they stare blankly, and sometimes do or say things that do not make sense
- They may also perform small twitching, stuttering, or fumbling actions while the seizure is ongoing.
- The child may experience odd feelings that signal the seizure is about to begin, but will likely not remember the events before or during the seizure itself.

UNKNOWN ONSET SEIZURES

When the beginning of a seizure is not known, it's now called an unknown onset seizure. A seizure could also be called an unknown onset if it's not witnessed or seen by anyone, for example when seizures happen at night or in a person who lives alone. As more information is learned, an unknown onset seizure may later be diagnosed as a focal or generalized seizure.

AURA

Some children have a strange sensation called an aura before a seizure. These sensations may include tingling, vision changes, anxiety, smelling an odor that is not there, or feeling ill.

THINKING AND PROBLEM SOLVING SKILLS

Some children with seizure disorders may have thinking and problem solving (knowing how to make decisions) skills at a lower level than children without seizures. This may happen for several different reasons. First, prolonged seizures damage important areas in the brain that help the child learn and problem solve. Also, some of the medications that children take (also known as antiepileptic or anticonvulsant drugs) can affect a child's memory or thinking abilities. These medications can also make it difficult for the child to pay attention.

FATIGUE OR SLEEP DISTURBANCES

Seizures can also affect a child's sleep, which may lead to daytime fatigue and concentration problems. The child or parent is often not aware that seizures are occurring at night. Medications prescribed to treat seizure disorders can also disturb sleep. This can be troublesome because lack of sleep is a trigger for seizures. For these children, it is very important to get an adequate amount of sleep on a nightly basis. Talk to your doctor if the child is not getting at least 8 hours of sleep per night.

DIAGNOSIS

Generally, a child is diagnosed with a seizure disorder after having two or more seizures. For this reason, it is important to talk with the child's pediatrician about any seizure symptoms like unusual daydreaming, fainting, shaking, muscle tremors, or convulsions. If your pediatrician suspects that a child has a seizure disorder, they may refer you to a doctor who specializes in the brain. This doctor is called a neurologist. They may also order additional tests including blood work, imaging of the brain, or a test of the electrical activity in the brain called an electroencephalogram (EEG). These tests will help the neurologist determine the cause and type of the seizures

Sometimes people have seizures that are not caused by the brain. Imbalances in the body that cause seizures include high fever, low blood sugar, drug use or withdrawal, or a problem with the electrolytes in someone's blood. These are not diagnosed as seizure disorders or epilepsy because they are related to an imbalance in the body that can be corrected.

MANAGEMENT

DURING A SEIZURE

- Keep calm
- Do not hold the child down or try to stop his/her movements
- Clear the area around the child of anything hard or sharp
- Loosen ties or anything around the neck that may make breathing difficult
- Put something flat and soft, like a folded jacket under the head
- If able, turn the child onto one side, to help keep the airway clear and prevent choking
- Remember- the child will not swallow his or her tongue! Do not try to force the mouth open with any object, especially fingers.
- Do not attempt CPR, except in the unlikely event that the heart stops beating and the child stops breathing after the seizure
- If possible, time the seizure and stay with the child until the seizure ends naturally
- As consciousness returns, provide comfort and reassurance to the child
- If your doctor has provided you with a medication, such as rectal Ativan to be administered during a seizure, do this now.

WHEN TO CALL 911

- The seizure lasts longer than 5 minutes
- The child has diabetes, a brain infection, high fever, heat exhaustion, or head injury
- The seizure happens in water
- The child stops breathing or turns blue
- A second seizure starts after the first ended or consciousness does not return

GENERAL SAFETY

Generally children with seizure disorders are more susceptible to falls and injury. It is important to keep them in a safe environment in case a seizure starts suddenly. In their home sharp corners should be padded, carpets and padding should be put on floors, and general child-proofing precautions such as a gate in front of the stairs and outlet covers should be in place. To prevent burns any heated appliance like an iron or curling iron should have an automatic shut off switch and be kept out of reach. If there is a fireplace in the home, install guards and close fireplace screens while a fire is burning. Chairs with arms are recommended to decrease risk of falling during a seizure. Also, the child should avoid top bunks. A lower bunk, a regular bed, a futon or even a mattress on the floor is a safer place to sleep for a child with seizures. Place a list of first-aid steps on the refrigerator or some other place where it is easy to find. Also, to best monitor the child, a baby monitor can be placed in the child's bedroom to alert you to the sound of a typical seizure.

Safety precautions for these children include wearing a life vest when near water, including backyard pools. Monitor the child at all times when near or in the water. It is also important to ensure that children wear a helmet during bicycling, rollerblading, or when using any other toys with wheels. A special helmet with a face guard may protect against head and facial injuries in children who have frequent severe seizures. Discuss with your doctor or healthcare provider whether helmet use is appropriate for your child.

It's important that people around the child and involved in their caretaking know what to do in case of a seizure. Children should wear an I.D. bracelet that states that they have seizures. When leaving the child with a babysitter, go over first-aid steps, write down the phone number where you or a relative can be reached, and give them a copy of this chapter. If your child is going to sleep at a friend or relative's house overnight, make that an adult in the house knows what to expect and what to do in the event of a seizure.

BATHROOM SAFETY

The bathroom is a particularly high risk area for children with seizures. Since a child having a seizure while bathing is at an increased risk of drowning, it is extremely important that they do so under close supervision. Keeping water levels low while the child bathes is also a good way to reduce their risk of drowning while having a seizure. Keep bath water temperatures low in order to avoid the risk of being burnt by hot water while having a seizure. If the child falls frequently during seizures, consider using a shower or bathtub seat with a safety strap. The caseworker can help you get this. Also, consider installing a hand-held shower nozzle so the child may be seated in the bathtub or shower while bathing. Keep bath water temperatures low in order to avoid the risk of being burnt by hot water while having a seizure. Finally, place extra padding under bathroom carpeting to decrease risk of injury. Finally, Hang bathroom doors so they open outwards instead of inwards. This will ensure that someone can still get to them, even if they fall in the doorway while having a seizure.

SCHOOL SAFETY

It is important to communicate with staff at the child's school to ensure they are safe and receiving educational support for their seizure disorder. Inform the school principal and school nurse that the child has a seizure disorder, and give them as much detail as possible about what their seizures look like, their medications, what triggers their seizures, and what to do when the child has a seizure. Provide school with all equipment necessary for your child's medical management (such as medications, a log to write down when medication was taken, etc, or other anything else you believe would benefit your child. It is also important that your child have an Individualized Education Plan (IEP) in place to make sure your child is receiving special support and education to meet their needs.

TREATMENT

There are a variety of treatments available for controlling seizures. It is important to discuss treatment options with the child's healthcare provider in order to determine which method best suits the child's needs.

AVOIDING TRIGGERS

Sometimes children with seizure disorders have triggers. Triggers are something that makes a child more likely to have a seizure. Triggers should be avoided to prevent seizures. Although each child is different, common triggers are things like lack of sleep, high stress, illness, missed medications, flashing lights, or substances like caffeine, alcohol, and drugs. Another very common trigger is lack of sleep. It's important to provide a consistent sleep schedule and ensure that they get at least 8 hours of sleep a night to prevent seizure activity. Caregivers should work together to identify and minimize the child's exposure to their seizure triggers.

ANTICONVULSANTS

Specific medications, known as anticonvulsants or antiepileptic drugs, are commonly prescribed to children with seizure disorders. Medications are the most common way that seizures are managed. It is very important that children prescribed these medications take them as directed by their doctor and on time. Sometimes these medications also make children groggy or upset their stomachs. These are important details to mention to the child's pediatrician. Some seizure medications also may affect the child's bones. Also ask your child's doctor if the child needs vitamin supplements.

OTHER TREATMENTS

Depending on how frequent the seizures occur, the pediatrician might mention other possible treatments such as diet changes, nerve stimulation, or even surgery. These treatments should be closely supervised by a doctor, and it is important to discuss the details and effects of each option with the child's pediatrician or neurologist. It is also important to eliminate any seizure triggers from the child's environment and generally decrease stress to prevent seizures.

PREVENTION

It is not possible to prevent most seizure disorders. However, it is possible to prevent seizures by making sure your child takes their medication on time and reporting any seizure activity to the child's neurologist. It is also possible to prevent injuries from occurring during a seizure. Follow the below steps to keep your child safe.

DOCTOR VISIT

Children with seizure disorders require frequent doctor visits. See Navigating a Doctor's Visit (pages 9 through 12) in the Introduction for information about how to prepare for a doctor visit.

It is important for your child to receive regular visits from their primary care doctor and pediatric neurologist. To find a pediatric neurologist located near you, visit the Epilepsy Foundation of America website at www.familyvoicesillinois.org/wp-content/uploads/2012/07/Neuro-Ep-Referral-List-2010.pdf.

COPING & SUPPORT

It is important to know that chronic health conditions can have a powerful effect on how children think about themselves and their ability to relate to others. Part of supporting a child with cerebral palsy is helping the child learn how to handle the stressors of their illness. The caregiver can help the child manage the stressors by encouraging the child to talk about how they feel about themselves and how they think others see them. It is important for caregivers to listen to and support the child about any anger or frustration. Caregivers should give positive feedback to the child regularly and celebrate their personal successes, even if they are different from other children their age. If the caregiver feels the child has low self-esteem, it's important to get professional help. Caseworkers, doctors, and other health care professionals can work to uncover underlying issues that are preventing the child from feeling good and work to improve the child's mental health. Churches, community centers, and local support groups are great sources of support and information. Below are some helpful organizations:

Epilepsy Foundation Of America

Landover, MD
Phone: 800-332-1000
www.epilepsy.com

Epilepsy Foundation Of Greater Chicago

Chicago, IL
Phone: 800-273-6027
www.epilepsychicago.org

American Epilepsy Society

Chicago, IL
Phone: 312-883-3800
www.aesnet.org

If you need assistance after hours, contact the 24 hour Epilepsy Foundation Support Hotline: 1 (800) 332-1000. For more information, contact your caseworker, healthcare provider, or the Epilepsy Foundation.

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CHAPTER SEVEN: SICKLE CELL DISEASE

OVERVIEW

Sickle cell disease (SCD) is a genetic disorder that affects the red blood cells. In healthy people, red blood cells are round, move easily through blood vessels, and carry oxygen to all parts of the body. In people with SCD, red blood cells change shape or “sickle” during times of physical or emotional stress. Sickle red blood cells are crescent or C-shaped, hard, and sticky. Sickle cells tend to clump together and block blood vessels, impeding blood flow and delivery of oxygen throughout the body. The poor oxygenation damages tissues and organs throughout the body, such as the spleen, brain, eyes, lungs, liver, heart, kidneys, penis, joints, bones, or skin.

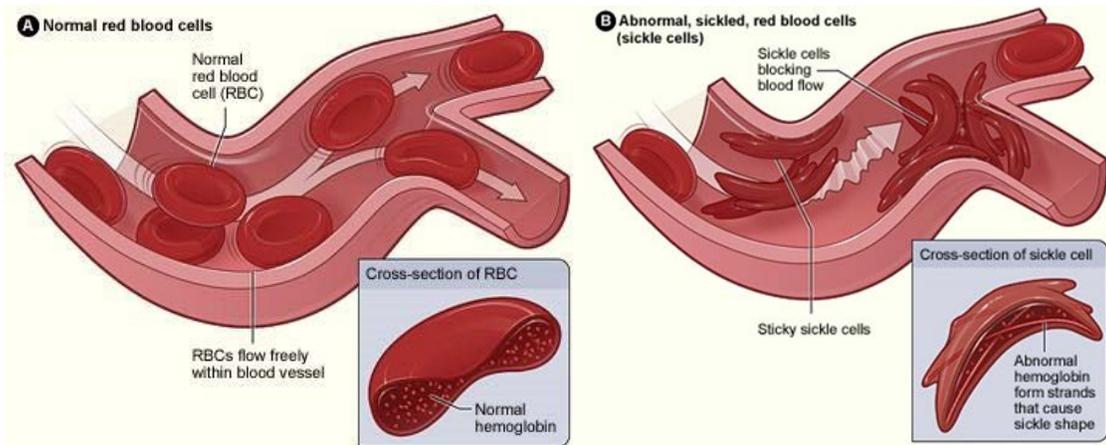


Figure 7.1 Figure A shows normal red blood cells flowing freely in a blood vessel. The inset image shows a cross-section of a normal red blood cell with normal hemoglobin. Figure B shows abnormal, sickled red blood cells blocking blood flow in a blood vessel. The inset image shows a cross-section of a sickle cell with abnormal (sickle) hemoglobin forming abnormal stiff rods (National Heart Lung and Blood Institute, 2016).

CAUSE

People have two copies of most genes, one copy from their father and one copy from their mother. Sometimes mistakes or defects occur in genes and cause genetic disorders. Genetic disorders are passed down in genes from parents to children. They are not contagious and cannot be “caught,” like a cold or flu.

SCD is a genetic disorder caused by a defect in the hemoglobin (HBB) gene. Hemoglobin is the molecule in red blood cells that delivers oxygen to cells throughout the body.

SCD is a recessive disorder, which means a person must inherit two defective copies of the HBB gene to have SCD. If only one defective copy of the HBB gene is inherited, the person will have sickle cell trait (SCT). People who have SCT do not have SCD, but can pass the defective HBB gene onto future children. SCT will not develop into SCD. If two normal functioning copies of the HBB gene are inherited, the person will not have SCD and cannot pass SCD onto future children.

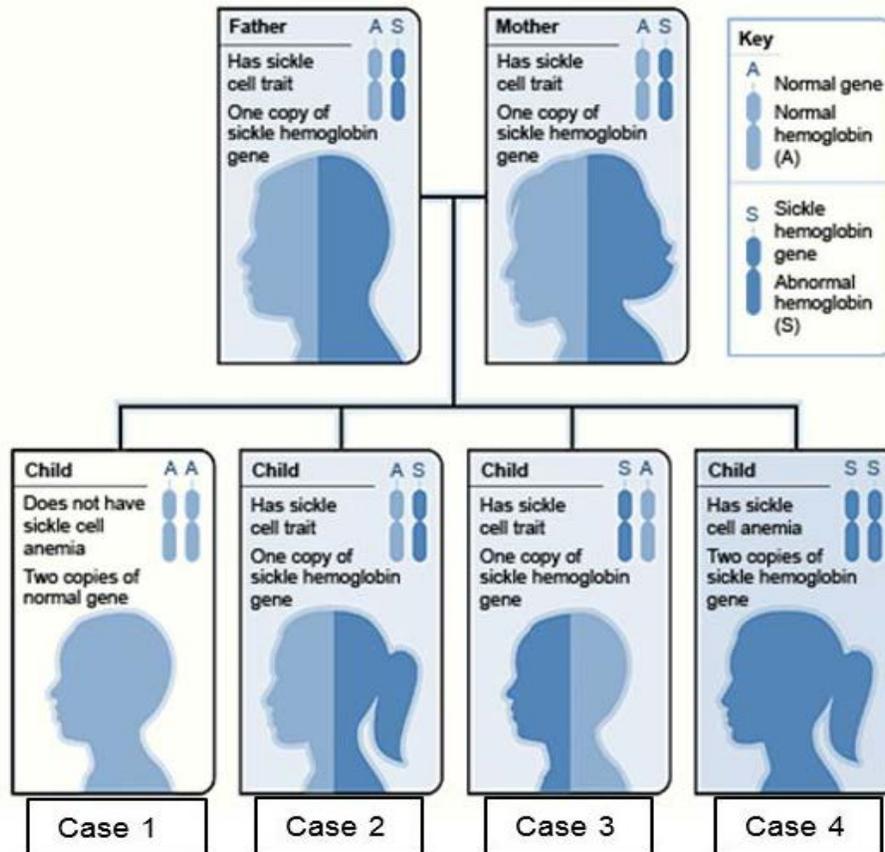


Figure 7.2 The image shows how sickle hemoglobin genes are inherited. A person inherits two hemoglobin genes—one from each parent. A normal gene will make normal hemoglobin (A). A sickle hemoglobin gene will make abnormal hemoglobin (S) (National Heart Lung and Blood Institute, 2016).

There are multiple types of SCD. The type of SCD is determined by the type of abnormal hemoglobin inherited from each parent. Normal hemoglobin is hemoglobin A; abnormal types of hemoglobin are hemoglobin S, C, D, E and thalassemia. The two most common types of SCD are HbSS and HbSC. People who inherit two genes for hemoglobin S have type HbSS. This form of the disease is commonly called sickle cell anemia and is usually the most severe form of the disease. People who inherit one gene for hemoglobin S and one gene for hemoglobin C have type HbSC. This form of the disease is usually a milder form of SCD.

SIGNS & SYMPTOMS

People with sickle cell trait (SCT) usually do not have any signs of the disease and live a normal life. However, medical complications have been reported in people with SCT during times of extreme stress and exertion, such as surgery, high altitudes, extremely hot or cold temperatures, severe dehydration, scuba diving, mountain climbing, and strenuous sports activities. People with SCT require special attention when participating in activities of extreme stress or exertion and should see a SCD specialist before participating. A list of local sickle cell clinics can be found below in the Doctor Visit section.

Signs and symptoms of sickle cell disease (SCD) vary from person to person. Typically, people with SCD start to develop signs and symptoms of the disease during the first year of life. Infants usually don't show any signs or symptoms of SCD at birth because fetal or baby hemoglobin protects the red blood

cells from sickling. When the infant is around five months of age, the fetal or baby hemoglobin is replaced by sickle hemoglobin and the cells begin to sickle. The most common signs and symptoms of SCD are described below.

ANEMIA

Sickle cells do not live as long as normal red blood cells. Normal red blood cells live about 90 to 120 days, but sickle cells only live about 10 to 20 days. The body is always making new red blood cells to replace the old cells; however, in SCD, the body cannot keep up with how fast the sickle cells are dying. Because of this, people with SCD usually have a lower than normal amount of red blood cells, which is called anemia. Anemia can cause tiredness, irritability, dizziness, a fast heart rate, and pale skin color.

Aplastic anemia is severe type of anemia. It occurs when an infection temporarily stops the body from making new blood cells leading to very low levels of blood.

INFECTION

The spleen is an organ in the abdomen that helps protect the body against infection. The spleen is responsible for filtering out germs and old blood cells. It also produces antibodies that attack bacteria. In SCD, the sickle cells damage the spleen and weaken its ability to protect the body against infection. Because of this, people with SCD are more at risk for infections. Infections can progress quickly and are a major cause of death in people with SCD. Fever is frequently the first sign of an infection. If the child's temperature is 100.4 °F or above, take the child to the emergency room right away. Do not attempt to treat a fever at home.

PAIN EPISODE OR VASO-OCCLUSIVE CRISIS (VOC)

Pain occurs when sickled cells get stuck in blood vessels, clog blood flow, and prevent oxygen from being delivered throughout the body. The pain can start suddenly, range from mild to severe, and last for any length of time. Most children with SCD are pain free between crises, but adolescents and adults may also suffer with chronic ongoing pain. Pain is the most common complication of SCD and is the most common reason people with SCD go to the emergency room.

STROKE

A stroke occurs when sickle cells get trapped in blood vessels and block blood flow to the brain. When this happens, brain cells are deprived of oxygen and begin to die. A stroke can cause learning problems, lifelong disabilities, and death. The common signs of a stroke are face drooping, arm weakness, and slurred speech. If you observe any of these signs, call 9-1-1 immediately. Use the acronym FAST to remember the warning signs of a stroke:

FACE: Ask the person to smile. Does one side of the face droop?

ARMS: Ask the person to raise both arms. Does one arm drift downward?

SPEECH: Ask the person to repeat a simple phrase. Is their speech slurred or strange?

TIME: If you observe any of these signs, call 9-1-1 immediately.

SPLENIC SEQUESTRATION

The spleen is a small organ, normally about the size of your fist. Splenic sequestration occurs when a large number of sickle red blood cells get trapped in the spleen, causing the spleen to suddenly enlarge. If too many red blood cells get trapped in the spleen, there will not be enough red blood cells available

to travel throughout the body and deliver oxygen to other vital organs. Symptoms include sudden weakness, pale lips, fast breathing, abdominal pain on the left side of body, enlarged abdomen, extreme thirst, and fast heartbeat. Parents of a child with SCD should learn how to feel and measure the size of their child's spleen and seek help if the spleen is enlarged or the abdomen is painful to touch. Splenic sequestration is a medical emergency, seek medical help immediately.

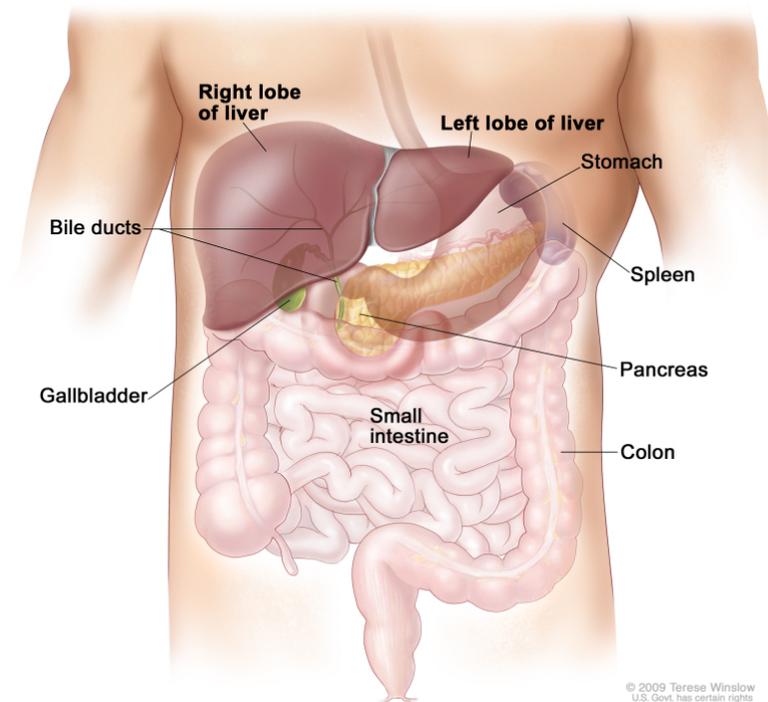


Figure 7.3 The spleen is an organ that is part of the lymphatic system. The spleen makes lymphocytes, filters the blood, stores blood cells, and destroys old blood cells. It is located on the left side of the abdomen near the stomach (U.S. National Library of Medicine, 2009)

ACUTE CHEST SYNDROME (ACS)

Acute chest syndrome occurs when sickle cells or bacteria gets trapped in the tissue of the lungs. Signs and symptoms include chest pain, coughing, difficulty breathing, and fever. ACS is a medical emergency, seek medical help immediately.

PRIAPISM

Priapism is a prolonged, unwanted, painful erection lasting more than two hours. It occurs when sickle cells become trapped in the erectile tissue of the penis. If an erection lasts more than four hours, seek medical help immediately.

DIAGNOSIS

SCD can be diagnosed before birth with two prenatal genetic tests, amniocentesis and chorionic villus (CVS). In amniocentesis, a doctor inserts a hollow needle through the abdominal wall into the uterus and removes a small amount of fluid from the sac around the baby. The fluid is tested to see whether both of the baby's HBB genes are normal. In CVS, a doctor threads a thin tube through the vagina and cervix

to the placenta and removes a tissue sample from the placenta using gentle suction. The tissue is tested to see whether both of the baby's HBB genes are normal.

SCD can be diagnosed after birth with a simple blood test. The blood is tested to see what type of hemoglobin is present.

Early diagnosis of SCD is important. In the United States, all states are required to test for SCD as part of the newborn screening. It is recommended that caseworkers and caregivers obtain a copy of the newborn screening for the child's home medical record.

MANAGEMENT

There is no single best treatment for all people with SCD. Treatment options are different for each person depending on the symptoms.

ANEMIA

The two main treatments for sickle cell anemia are oral folic acid supplements and blood transfusions.

FOLIC ACID

Folate is a vitamin the body needs to make new red blood cells. In SCD, folate stores are often depleted because the body is working over-time in an attempt to make enough new red blood cells. Folic acid replenishes the depleted folate stores.

BLOOD TRANSFUSION

A blood transfusion is a procedure that takes healthy blood from one person and puts it into someone who does not have enough blood or does not have enough healthy blood. In severe anemia, blood transfusions are used to increase red blood cell counts. It is a safe, common procedure.

INFECTION

There are many ways caregivers can prevent or reduce the number of infections, including using proper handwashing technique, keeping the child and family members up to date with all immunizations, and administering prescribed antibiotics.

HANDWASHING

Handwashing is the number one way to prevent the spread of germs.

IMMUNIZATIONS

All members of the household should be up to date with all recommended childhood vaccines, plus influenza, pneumococcal, and meningococcal vaccines.

ANTIBIOTICS

Antibiotics kill or stop the growth of bacteria and are used to prevent infections. Take penicillin, or other antibiotic prescribed by a doctor, every day until at least 5 years old.

PAIN EPISODE

Pain episodes can be avoided by preventing red blood cells from becoming sickle-shaped. Ways to help sickle cells stay round include:

PREVENTION

- Drinking plenty of fluids. Ask your primary care doctor what amount is right for your child's age.
- Avoiding extremely hot or cold temperatures. Any sports-related injuries should NOT be treated with ice.
- Avoiding places or situations with low oxygen, such as high altitudes, strenuous athletic training, or over exertion.
- Getting plenty of rest and taking frequent breaks during exercise.

TREATMENT

Severe pain, unusual pain, or pain that won't go away should be treated in a hospital. Mild pain can be treated at home with over the counter pain relievers, such as Advil (ibuprofen) or Tylenol (acetaminophen), plenty of fluids, and rest. NEVER administer aspirin to a child. Aspirin use in children is associated with Reyes Syndrome, which can cause life-threatening swelling in the liver and brain.

Over the counter pain relievers can mask a fever, so it is important to ALWAYS take the child's temperature before administering. Remember, a fever is a medical emergency in people with sickle cell disease. If the child's temperature is 100.4 °F or above, take the child to the emergency room right away. Do not attempt to treat fever at home.

STROKE

Not all children with SCD are at equal risk for stroke. A special procedure, called transcranial Doppler ultrasound (TCD), is used to identify children who are at risk for a stroke. If a child is found to be at risk for a stroke, a doctor might recommend monthly blood transfusions to help prevent a stroke. A blood transfusion is a procedure that takes healthy blood from one person and puts it into someone who does not have enough blood or does not have enough healthy blood. In stroke prevention, blood transfusions are used to reduce the proportion of sickle red blood cells by increasing the number of healthy red blood cells. It is a safe, common procedure.

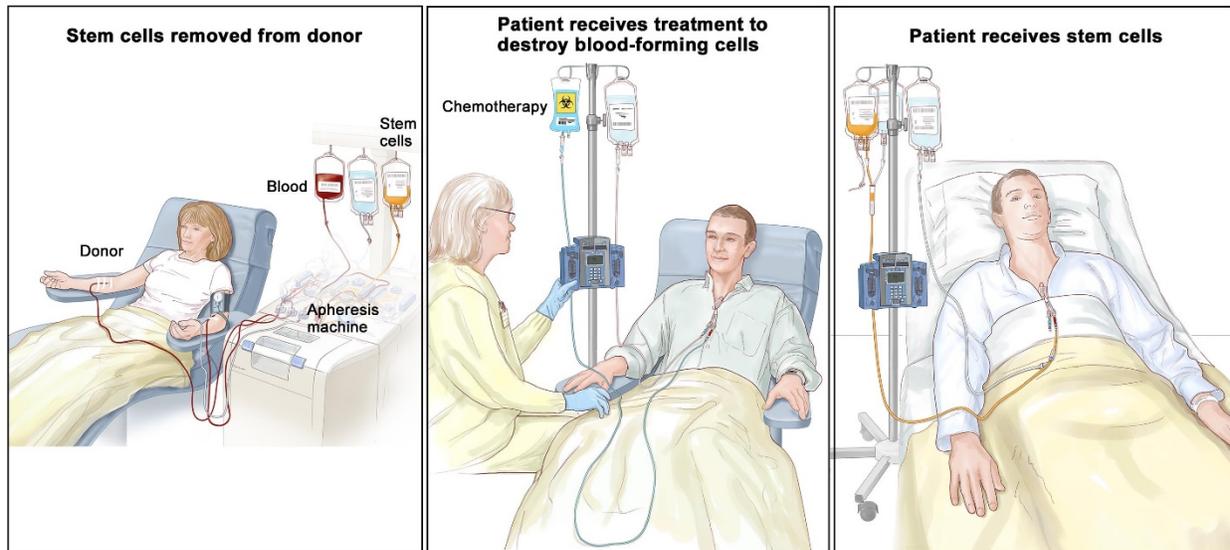
SPLenic SEQUESTRATION

Blood Transfusion. A blood transfusion is a procedure that takes healthy blood cells from one person and puts it into someone who does not have enough blood cells or does not have enough healthy blood. In severe splenic sequestration, blood transfusions are used to increase the amount of red blood cells available to travel throughout the body and deliver oxygen. It is a safe, common procedure.

Splenectomy. A splenectomy may be performed if frequent splenic sequestration episodes occur. Splenectomy is the surgical removal of the spleen.

HEMATOPOIETIC STEM CELL TRANSPLANTATION (HSCT)

The only known cure for SCD is a bone marrow transplant, also known as hematopoietic stem cell transplant (HSCT). Bone marrow is a soft, fatty tissue inside the center of the bones where red blood cells are made. A bone marrow transplant is a procedure that takes healthy bone marrow cells from one person (donor) and puts them into someone whose bone marrow is not working properly (recipient).



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Figure 7. 4 Stem cell transplant. (Step 1): Blood is taken from a vein in the arm of the donor. The blood flows through a machine that removes the stem cells. (Step 2): The recipient receives chemotherapy to kill blood-forming cells. (Step 3): The recipient receives stem cells through a catheter placed into a blood vessel in the chest. (National Cancer Institute, 2011)

A well-matched donor is needed to have the best chance for a successful transplant. After a bone marrow transplant, the bone marrow of a person with SCD should start producing healthy red blood cells, instead of sickle cells. However, bone marrow transplants are only used in cases of severe SCD because they are very risky and can have serious side effects, including death. In addition, most people with SCD are either too old for a transplant or don't have a relative who is a good enough genetic match for them to act as a donor.

HYDROXYUREA

For individuals that are not good candidates for a bone marrow transplant, Hydroxyurea can reduce symptoms and prolong life. Hydroxyurea is the only drug that the Food and Drug Administration (FDA) has approved for the management of SCD. Hydroxyurea prevents the complications of SCD by increasing fetal hemoglobin (HbF), which stops red blood cells from sickling. Hydroxyurea can have serious side effects, like bone marrow failure, and, thus, is only used in patients who have had at least three pain crises in 12 months. Blood tests are required every four to eight weeks while taking hydroxyurea to measure white blood cell, red blood cell, and platelet counts, and ensure the bone marrow is functioning appropriately.

PREVENTION

You cannot prevent sickle cell disease. However, you can follow the steps mentioned in the management section to control the disease and prevent its symptoms.

DOCTOR VISIT

Children with SCD require frequent doctor visits. See Navigating a Doctor's Visit (pages 9 through 12) in the Introduction for information about how to prepare for a doctor visit.

Care plans will vary from child to child. Some children will need to visit the doctor more often than others. In addition to routine primary care doctor visits, children with SCD should also visit a SCD specialist, genetics counselor, and vision/hearing screenings.

SCD SPECIALIST – EVERY 3 TO 12 MONTHS

The National Heart, Lung, and Blood Institute recommends children with SCD or SCT visit a SCD specialist every three to 12 months. The frequency of visits will vary on the severity of the disease.

Below is a list of local sickle cell centers. If a sickle cell center is not located near your home, contact your primary care provider to find a local doctor that specializes in blood disorders.

Advocate Children’s Hospital – Keyser Center

Oak Lawn, IL

Phone: 708-684-3898

www.advocatechildrenshospital.com/ach-sickle-cell-disease

University Of Chicago Pediatric Specialists At La Rabida Children’s Hospital

Chicago, IL

Phone: 773-256-5759

www.larabida.org/page-sickle-cell

University Of Illinois Hospital & Health System

Chicago, IL

Phone: 312-413-8666

hospital.uillinois.edu/primary-and-specialty-care/sickle-cell

St. Louis Children’s Hospital

St. Louis, MO

Phone: 314-454-5437

www.stlouischildrens.org/our-services/sickle-cell-disease-program

Ann & Robert H. Lurie Children’s Hospital

Chicago, IL

Phone: 800-543-7362

www.luriechildrens.org/en-us/care-services/specialties-services/hematology-oncology-stem-cell-transplantation/programs/pages/sickle-cell-disease-program.aspx

GENETICS COUNSELOR – AT PUBERTY

It is recommended that children with SCD or SCT receive genetics counseling when they reach puberty or sexual maturity. Children with SCD or SCT can pass the gene for SCD onto their children. A genetics counselor will perform genetic testing to determine their risk for having a child with SCD. A genetics counselor can also answer any questions and explain the choices that are available. If you do not already have a genetics counselor, contact your primary care doctor or SCD specialist for a referral.

VISION – EVERY YEAR

Vision and hearing screenings should be performed annually. Sickle cells can block blood flow in the blood vessels in the eye and can cause blindness, also known as sickle cell retinopathy.

COPING & SUPPORT

It is important to know that chronic health conditions can have a powerful effect on how children think about themselves and their ability to relate to others. Encouraging children to talk about how they feel about themselves and how they think others see them can help, both emotionally and socially. If you feel the child has low self-esteem, professional help can be sought. Caseworkers, doctors, and other health care professionals can work to uncover underlying issues that are causing low self-esteem.

In addition, community centers, SCD clinics, and SCD organizations are great sources of support and information for both the child and caregiver. Below are some helpful SCD organizations:

American Sickle Cell Anemia Association

www.ascaa.org

Sickle Cell Information Center

www.scinfo.org

Sickle Cell Disease Association of Illinois

www.sicklecelldisease-illinois.org

Have a Heart for Sickle Cell Anemia Foundation

www.hhsca.org

Sickle Cell Disease Association of America

www.sicklecelldisease.org

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- National Stroke Association. (2017). Act FAST. Retrieved from <http://www.stroke.org/understand-stroke/recognizing-stroke/act-fast>
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- U.S. National Library of Medicine. (2009). Spleen. Retrieved from <https://www.ncbi.nlm.nih.gov/pubmedhealth/PMHT0022189/>